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**Assessing children's and teenagers' bereavement when a sibling dies from cancer: a secondary analysis.**

Birenbaum LK.

*Child Care Health Dev 2000;26(5):381-400.*

The purposes of this paper are to: (1) provide conceptual and empirical descriptions of the most frequently reported children's and teenagers' responses in anticipation of and after the death of a sibling from cancer; (2) describe these behaviours by age groups (3-5, 6-11 and 12-19 years); and (3) discuss the clinical utility of two new scales measuring children's bereavement. The literature review and an exploratory secondary analysis from a prospective longitudinal design were used to develop the empirical criteria for behavioural items indicative of bereavement. Data from four points in time: (1) 2 months before a child's death; (2) 2 weeks after death; (3) 4 months after death; and (4) 12 months after death were treated as a cross-sectional design, because of sample size, to describe bereavement behaviours and to discuss the beginning development of a screening tool for childhood bereavement services.


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**The measurement of symptoms in children with cancer.**


The purpose of this study was to determine symptom prevalence, characteristics, and distress in children with cancer. The Memorial Symptom Assessment Scale (MSAS) 10-18, a 30-item patient-rated instrument adapted from a previously validated adult version, provided multidimensional information about the symptoms experienced by children with cancer. This instrument was administered to 160 children with cancer aged 10-18 (45 inpatients, 115 outpatients). To confirm the instrument's reliability and validity, additional data about symptoms were collected from both the parents and the medical charts, and retesting was performed on a subgroup of inpatients. Patients could easily complete the scale in a mean of 11 minutes. The analyses supported the reliability and validity of the MSAS 10-18 subscale scores as measures of physical, psychological, and global symptom distress, respectively. Symptom prevalence ranged from 49.7% for lack of energy to 6.3% for problems with urination. The mean (+/- SD) number of symptoms per inpatient was 12.7 +/- 4.9 (range, 4-26), significantly more than the mean 6.5 +/- 5.7 (range, 0-28) symptoms per outpatient. Patients who had recently received chemotherapy had significantly more symptoms than patients who had not received chemotherapy for more than 4 months (11.6 +/- 6.0 vs. 5.2 +/- 5.1), and those patients with solid tumors had significantly more symptoms than patients with either leukemia, lymphoma, or central nervous system malignancies (9.9 +/- 7.0 vs. 6.8 +/- 5.5 vs. 6.8 +/- 5.0 vs. 8.0 +/- 6.1). The most common symptoms (prevalence > 35%) were lack of energy, pain, drowsiness, nausea, cough, lack of appetite, and psychological symptoms (feeling sad, feeling nervous, worrying, feeling irritable). Of the symptoms with prevalence rates > 35%, those that caused high distress in more than one-third of patients were feeling sad, pain, nausea, lack of appetite, and...
feeling irritable. Subscale scores demonstrated large variability in symptom distress and could identify subgroups with high distress. The prevalence, characteristics, and distress associated with physical and psychological symptoms could be quantified in older children with cancer. The data confirm a high prevalence of symptoms overall and the existence of subgroups with high distress associated with one or multiple symptoms. Symptom distress is relatively higher among inpatients, children with solid tumors, and children who are undergoing antineoplastic treatment. Systematic symptom assessment may be useful in future epidemiological studies of symptoms and in clinical chemotherapeutic trials. Symptom epidemiology may also provide a focus for future clinical trials related to symptom management in children with cancer.


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Symptoms and suffering at the end of life in children with cancer.

Deremo DE, Fahner JB.


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Symptoms and suffering at the end of life in children with cancer.

Goldman A.


No abstract available

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All the king's horses and all the king's men: pain management from hospital to home.

Hellsten MB.


Pain management in children with cancer continues to be less than adequate in most settings. Children in pain often have multiple caregivers, each imposing a significant risk to adequate pain management if they are uninformed about the principles of pain management and the child's specific pain management regimen. This article reviews current pain management guidelines for children, obstacles to adequate pain management, and the challenges of managing pain across care settings (hospital to home). Using the hospice model of care, case studies are presented that illustrate both threats to adequate pain control and the coordination of care necessary to ensure consistent pain management between the hospital and home. Implications for improvement in pain management between settings and future directions for advancing skills and knowledge related to pain management in children with cancer are provided.

Parental cigarette smoking, hard liquor consumption and the risk of childhood brain tumors–a case-control study in northeast China.

Hu J, Mao Y, Ugnat AM.


In this study we examine the effect of parents' lifestyles on the risk of childhood brain tumors. Parents of 82 children newly diagnosed with primary malignant brain tumors and 246 individually matched hospital controls were interviewed in the hospital wards between September 1991 and December 1996. Data were collected on socioeconomic status, parental lifestyle prior to and during the pregnancy, and family history. Odds ratios and 95% confidence intervals were derived through conditional logistic regression. The risk of childhood brain tumors was associated with paternal use of hard liquor prior to the pregnancy: the odds ratios were 3.72 (95% CI = 1.91-7.26) for < or = 15 years of hard liquor consumption and 4.06 (95% CI = 1.09-15.21) for > or = 16 years of hard liquor consumption compared with never consuming hard liquor (test for trend p = 0.0001); the odds ratios increased with increasing lifetime hard liquor consumption. There is little evidence to support an association between childhood brain tumors and parents' smoking prior to or during pregnancy.


Care of children who are dying of cancer.

Morgan ER, Murphy SB.


Care of terminally ill children with cancer.


Symptoms and suffering at the end of life in children with cancer.

Sandoval C.

Symptoms and suffering at the end of life in children with cancer.


Background: Cancer is the second leading cause of death in children, after accidents. Little is known, however, about the symptoms and suffering at the end of life in children with cancer. Methods: In 1997 and 1998, we interviewed the parents of children who had died of cancer between 1990 and 1997 and who were cared for at Children's Hospital in Boston, the Dana-Farber Cancer Institute, or both. Additional data were obtained by reviewing medical records. Results: Of 165 eligible parents, we interviewed 103 (62 percent), 98 by telephone and 5 in person. The interviews were conducted a mean (+/-SD) of 3.1 +/-1.6 years after the death of the child. Almost 80 percent died of progressive disease, and the rest died of treatment-related complications. Forty-nine percent of the children died in the hospital; nearly half of these deaths occurred in the intensive care unit. According to the parents, 89 percent of the children suffered "a lot" or "a great deal" from at least one symptom in their last month of life, most commonly pain, fatigue, or dyspnea. Of the children who were treated for specific symptoms, treatment was successful in 27 percent of those with pain and 16 percent of those with dyspnea. On the basis of a review of the medical records, parents were significantly more likely than physicians to report that their child had fatigue, poor appetite, constipation, and diarrhea. Suffering from pain was more likely in children whose parents reported that the physician was not actively involved in providing end-of-life care (odds ratio, 2.6; 95 percent confidence interval, 1.0 to 6.7). Conclusions: Children who die of cancer receive aggressive treatment at the end of life. Many have substantial suffering in the last month of life, and attempts to control their symptoms are often unsuccessful. Greater attention must be paid to palliative care for children who are dying of cancer. (N Engl J Med 2000;342:326-33.) (C) 2000, Massachusetts Medical Society.

Understanding of prognosis among parents of children who died of cancer: impact on treatment goals and integration of palliative care.


CONTEXT: Parents' understanding of prognosis or decision making about palliative care for children who die of cancer is largely unknown. However, a more accurate understanding of prognosis could alter treatment goals and expectations and lead to more effective care. OBJECTIVES: To evaluate parental understanding of prognosis in children who die of cancer and to assess the association of this factor with treatment goals and the palliative care received by children. DESIGN, SETTING, AND PARTICIPANTS: Survey, conducted between September 1997 and August 1998, of 103 parents of children who received treatment at the Dana-Farber Cancer Institute and Children's Hospital, Boston, Mass, and who died of cancer between 1990 and 1997 (72% of those eligible and those located) and 42 pediatric oncologists. MAIN OUTCOME MEASURE: Timing of parental understanding that the child had no realistic chance for cure compared with the timing of physician understanding of this prognosis, as documented in the medical record. RESULTS: Parents first recognized that the child had no realistic chance for cure a mean (SD) of 106 (150) days before the child's death, while physician recognition occurred earlier at 206 (330) days before death. Among children who died of progressive disease, the group characterized by earlier recognition of this prognosis by both parents and physicians had earlier discussions of hospice care (odds ratio [OR], 1.03; 95% confidence interval [CI], 1.01-1.06; P = .01), better parental ratings of the quality of home care (OR, 3.31; 95% CI, 1.15-9.54; P = .03), earlier institution of a do-not-resuscitate order (OR, 1.03; 95% CI, 1.00-1.06; P = .02), less use of
cancer-directed therapy during the last month of life (OR, 2.80; 95% CI, 1.05-7.50; P = .04), and higher likelihood that the goal of cancer-directed therapy identified by both physician and parent was to lessen suffering (OR, 5.17; 95% CI, 1.86-14.4; P = .002 for physician and OR, 6.56; 95% CI, 1.54-27.86; P = .01 for parents). CONCLUSION: Considerable delay exists in parental recognition that children have no realistic chance for cure, but earlier recognition of this prognosis by both physicians and parents is associated with a stronger emphasis on treatment directed at lessening suffering and greater integration of palliative care. JAMA. 2000;284:2469-2475.


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An ecology of love: Aspects of music therapy in the pediatric oncology environment.

Aasgaard T.


This paper explores how music therapy can assist patients and relatives in the processes of making friendship and love audible in a child cancer ward. Four short patient histories are presented to illustrate a health-oriented, ecological music therapy practice. Two histories describe how texts, made by patients, become songs, and how the songs are performed and used. Another two histories deal with musical communication with dying children and their parents. The paper indicates that these interventions may involve more than palliation (making a disease less severe and unpleasant without removing its cause). Not least, such activities can make it possible for the sick child to expand from being "just a patient" into playing, if only for a moment, a more active social role. The processes of artistic interplay, in- and outside the sickroom, influence various relationships in the child's social environment.

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A "good death" revisited in the context of doctor-patient relationships.

Abratt RP.


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When does primary care turn into palliative care?

Back AL, Curtis JR.


No abstract available

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Syringe driver drug compatibility database and patient information leaflets on the Internet.

Back I.


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Tumor- & treatment-related side effects after multimodal therapy of childhood intracranial germ cell tumors

Benesch M, Lackner H, Schagerl S, Gallistl S, Frey EM, Urban C.


Multimodality treatment approaches have dramatically improved the outcome of patients with intracranial germ cell tumors and are resulting in an increasing number of long-term survivors. The aim of the present study was to evaluate prospectively the development of side effects in children, adolescents and young adults after treatment for intracranial germ cell tumors. Nine patients with a median age of 14 y at diagnosis and a median follow-up of 7.25 y underwent a detailed long-term evaluation including physical and neuro-ophthalmologic examinations, routine laboratory and endocrine stimulation tests, neuropsychometric testing, audiometry and spirometry at repeated intervals. Endocrine deficiencies requiring hormone replacement therapy occurred in all patients. Neuro-ophthalmologic side effects were observed in 8 of the 9 patients, urinary electrolyte wasting in 4 of the 9, alopecia in 3 of the 9 and high-frequency hearing loss in 2 of the 9. Neuropsychologic examinations revealed pathologic results in all five tested patients. CONCLUSION: The present study indicates that former intracranial germ cell tumor patients suffer from remarkable long-term side effects, and that some of these late effects can develop or worsen months or years after cessation of oncologic therapy. Since life quality is an important parameter of cancer survival, careful follow-up of long-term survivors is mandatory, aimed at counteracting side effects as early as possible and therefore at minimizing long-term morbidity, which may considerably compromise quality of life.


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Measuring pain accurately in children with cognitive impairments: refinement of a caregiver scale.
Breau LM, Camfield C, McGrath PJ, Rosmus C, Finley GA.


OBJECTIVE: To examine whether typical pain behavior, as reported by caregivers, could be used prospectively to predict future pain behavior and to derive a subset of core items from the Non-Communicating Children's Pain Checklist. STUDY DESIGN: Caregivers (n = 33) of children with cognitive impairments completed the Non-Communicating Children's Pain Checklist retrospectively and immediately after subsequent episodes of pain and distress in their homes. Odds ratios were computed for checklist items, and multiple regressions were used to predict numerical pain and distress ratings with items that had significant odds ratios. A logistic regression was used to test whether the items found to predict pain could correctly classify the presence or absence of pain in a new cohort of 63 children with similar cognitive impairments. RESULTS: Seven of the checklist items had significant odds ratios: Cranky, Seeking Comfort, Change in Eyes, Less Active, Gesture to Part That Hurts, Tears, and Gasping. This subset of items significantly predicted numerical pain ratings by caregivers (multiple R =.70), but not distress ratings (multiple R =.31). In a second group of 63 children with cognitive impairments, this subset of items displayed 85% sensitivity and 89% specificity for pain. CONCLUSION: A subset of items from the Non-Communicating Children's Pain Checklist could predict pain in children with cognitive impairments. Caregivers' retrospective reports may be useful for clinicians making judgments about pain in these children.

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Hospice care for the child with AIDS.

Buckingham RW, Meister EA.


Hospice care was established to provide palliative (i.e., noncurative) services for the dying and their families. The advent of the AIDS epidemic has posed a challenge to hospice care, particularly for the child dying of the disease, and has adapted to modified palliative services. Parents, with a child dying of AIDS, must deal with many issues of disclosing the disease status to the child, coping with the emotions of losing a child, and when and where to incorporate hospice services into the dying process. Optimizing home based *hospice* care involves; (1) Nutritional management, (2) Prevention of opportunistic infections, (3) Pain management, and (4) Protection of Non-HIV positive members of hospice care. For the dying, hospice strives to achieve a peaceful death and provide supportive intervention for the survivors. 2001 Elsevier Science Inc. All rights reserved

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End-of-life care in the pediatric intensive care unit: Attitudes and practices of pediatric critical care physicians and nurses.

Burns JP, Mitchell C, Griffith JL, Truog RD.


Objective: To determine the attitudes and practices of pediatric critical care attending physicians and pediatric critical care nurses on end-of-life care. Design: Cross- sectional survey. Setting: A random sample of clinicians at 31
pediatric hospitals in the United States. Measurements and Main Results: The survey was completed by 110/130 (85%) physicians and 92/130 (71%) nurses. The statement that withholding and withdrawing life support is unethical was not endorsed by any of the physicians or nurses. More physicians (78%) than nurses (57%) agreed or strongly agreed that withholding and withdrawing are ethically the same (p < .001). Physicians were more likely than nurses to report that families are well informed about the advantages and limitations of further therapy (99% vs. 89%; p < .003); that ethical issues are discussed well within the team (92% vs. 59%; p < .0003), and that ethical issues are discussed well with the family (91% vs. 79%; p < .0002). On multivariable analyses, fewer years of practice in pediatric critical care was the only clinician characteristic associated with attitudes on end-of-life care dissimilar to the consensus positions reached by national medical and nursing organizations on these issues. There was no association between clinician characteristics such as their political or religious affiliation, practice-related variables such as the size of their intensive care unit or the presence of residents and fellows, and particular attitudes about end-of-life care. Conclusions: Nearly two-thirds of pediatric critical care physicians and nurses express views on end-of-life care in strong agreement with consensus positions on these issues adopted by national professional organizations. Clinicians with fewer years of pediatric critical care practice are less likely to agree with this consensus. Compared with physicians, nurses are significantly less likely to agree that families are well informed and ethical issues are well discussed when assessing actual practice in their intensive care unit. More collaborative education and regular case review on bioethical issues are needed as part of standard practice in the intensive care unit.

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Is the Kasai operation still indicated in children older than 3 months diagnosed with biliary atresia?


OBJECTIVES: The prognosis of hepatopportoenterostomy (the Kasai operation) for biliary atresia worsens when the age at surgery increases. This study examines whether the Kasai operation remains justified after 3 months of life. STUDY DESIGN: Records for all patients with biliary atresia living in France and born in the years 1986 to 1996 were reviewed, and patients were classified into 3 groups: group 1 (n = 30), no contraindication to the Kasai operation, but orientation to de novo transplantation; group 2 (n = 380), age at Kasai operation <90 days; and group 3 (n = 60), age at Kasai operation > or =90 days. Survival with native liver, survival after liver transplantation, and overall survival (Kaplan-Meier method) were compared by using the log-rank test. RESULTS: Five-year (10-year) survival with native liver was 35% (30%) in group 2 and 25% (22%) in group 3 (P =.03). Five-year overall survival was 57%, 74%, and 55% in groups 1, 2, and 3, respectively (P =.003). Poor results in groups 1 and 3 were mainly due to increased pre-transplantation mortality, but survival after transplantation was not significantly different in the 3 groups. CONCLUSIONS: Performance of the Kasai operation after 3 months of age is justified in selected cases, because it may obviate liver transplantation. Preoperative evaluation should exclude patients with advanced liver disease for whom liver transplantation should not be delayed.

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**Congenital adrenal hyperplasia: management during critical illness.**

BACKGROUND: Little is known of the optimal dose and administration schedule of hydrocortisone in critically ill patients with congenital adrenal hyperplasia (CAH) caused by 21-hydroxylase deficiency. AIM: To determine plasma cortisol concentrations after intravenous administration of hydrocortisone in children with CAH and to relate these to plasma cortisol concentrations achieved by endogenous secretion in the stress of critical illness in previously healthy children. METHODS: Plasma cortisol concentrations were measured in 20 patients with classical CAH (median age 11.2 years, range 6.1-16.4) following intravenous administration of hydrocortisone 15 mg/m(2); and in 60 critically ill mechanically ventilated children (median age 2.5 years, range 0.25-16.3) on admission to the paediatric intensive care unit and for 24 hours thereafter. RESULTS: In the CAH patients, plasma cortisol reached a mean peak of 1648.3 nmol/l (SD 511.9) within 10 minutes of the intravenous bolus, and fell rapidly thereafter; levels remained greater than 450 nmol/l for 2.5 hours only. In critically ill children, mean plasma cortisol on admission to the intensive care unit was 727 nmol/l (SD 426.1). Cortisol concentrations remained raised during the first 24 hours. CONCLUSIONS: Critically ill patients with classical CAH may be best managed with a single intravenous hydrocortisone bolus followed by a constant rate infusion of hydrocortisone.

Evaluation of an oral care protocol intervention in the prevention of chemotherapy-induced oral mucositis in paediatric cancer patients.

Cheng KKF, Molassiotis A, Chang AM, Wai WC, Cheung SS.


Oral mucositis is the most frequent and severe complication of chemotherapy in children with cancer that can aggravate the child's clinical condition and increase the risk of infection. This prospective comparative study was designed to determine the effectiveness of a preventive oral care protocol in reducing chemotherapy-induced oral mucositis in children with cancer. During an 8-month period, 42 children aged 6 to 17 years with haematological malignancies or solid tumours were evaluated. The 21 children who were included in the first 4-month period of the study constituted the control group. Another 21 children were enrolled in the subsequent 4 months and were assigned to the experimental group, in which they were given an oral care protocol intervention. The oral care protocol consisted of tooth brushing, 0.2% chlorhexidine mouth rinse and 0.9% saline rinse. Children in both groups were evaluated twice a week for 3 weeks. The incidence of ulcerative lesions, severity of oral mucositis and the related pain intensity were used as the main outcome variables. A 38% reduction in the incidence of ulcerative mucositis was found in children using the oral care protocol compared with children in the control group. The severity of oral mucositis (P = 0.000002) and the related pain (P = 0.0001) were significantly reduced with the intervention. These results support the preventive use of the oral care protocol in paediatric cancer patients who undergo chemotherapy for cancer treatment. (C) 2001 Elsevier Science Ltd. All rights reserved.

Down's syndrome and acute lymphoblastic leukaemia: clinical features and response to treatment.

AIMS: To examine the clinical and biological features of acute lymphoblastic leukaemia in children with Down's syndrome (DS), to compare their survival with other children, and to determine if entry to trials and survival has improved. METHODS: Examination of presenting features and response to treatment in patients treated in two consecutive national trials, MRC UKALL X and XI. RESULTS: The proportion of children with DS was significantly higher in UKALL XI (1.9%) than UKALL X (0.9%). Children with DS tended to be under 10 years and to have the common ALL subtype. Cytogenetic analysis showed that favourable features, such as high hyperdiploidy and t(12;21) were less frequent but also that there was a lack of translocations associated with a poor prognosis. Children with DS showed no increase in risk of relapse at any site but their survival and event free survival were inferior to other children. These results were caused by an increased number of infective deaths during remission (11% compared to 2%). At five years overall survival was 73% in DS children compared with 82% in other children; event free survival was 53% compared to 63% in non-DS children. CONCLUSIONS: Entry of children with DS to national trials has increased and survival has improved. However they remain at risk of relapse and also of treatment related mortality. These findings emphasise the need for both intensive chemotherapy and optimal supportive care.

Response to influenza immunisation during treatment for cancer.

Chisholm JC, Devine T, Charlett A, Pinkerton CR, Zambon M.

AIMS: To assess the annual risk of influenza infection in children with cancer and the immunogenicity of a trivalent split virus influenza vaccine in these children. METHODS: Eighty four children with cancer were tested for susceptibility to the circulating strains of influenza virus in autumn 1995 and 1996. Non-immunised children were reassessed the following spring for serological evidence of natural infection. Forty two patients received two doses of influenza vaccine. These children were receiving continuing chemotherapy for acute lymphoblastic leukaemia or were within six months of completing chemotherapy. RESULTS: Among the 84 children tested for influenza virus susceptibility only 8% of patients were fully protected (antibody titres $\geq 40$) against all three of the prevalent influenza virus strains; 33% were susceptible to all three viruses. Evidence of acquired natural infection was seen in 30% of unimmunised patients. Among immunised susceptible patients, 66% made some protective response to the vaccine and 55% showed protective antibody titres to all three viral strains following vaccination. Older age was associated with increased response to the H1N1 and H3N2 vaccine components, but total white cell count or neutrophil count at immunisation, type of cancer, or length of time on treatment for acute lymphoblastic leukaemia did not affect response. CONCLUSIONS: Most children with cancer studied were at risk of influenza infection. A significant response to immunisation was seen, supporting annual influenza vaccination for children being treated for cancer.

Does continuity of care matter? Yes: consistent contact with a physician improves outcomes.
Christakis DA.


http://www.ewjm.com/cgi/content/full/175/1/4

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**Growth study of cri du chat syndrome.**

Collins MS, Eaton-Evans J.


We compared the growth of children with cri du chat (5p-) syndrome with the 1990 UK growth curves. Most subjects had impaired growth, particularly of head circumference. The more emaciated the child the more pronounced the microcephaly, showing the need for growth and nutrition monitoring.

http://www.archdischild.com/cgi/content/full/archdischild;85/4/337
http://www.archdischild.com/cgi/content/abstract/archdischild;85/4/337

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**Neuroblastoma: changing incidence and survival in young people aged 0-24 years. A report from the North of England Young Persons' Malignant Disease Registry.**

Cotterill SJ, Parker L, More L, Craft AW.


**BACKGROUND AND PROCEDURE:** Population based data for neuroblastoma in children and young adults under 25 years at diagnosis were ascertained from the Northern Region Young Persons' Malignant Disease Registry for the period 1968-1995. Age-standardised incidence rates were calculated (ASR) and changes in incidence and survival were investigated. Over the study period 144 patients were registered, of these 136 were children under 15 years at diagnosis (median age: 2.2 years, ASR: 8.6 cases per million children per year), and 8 were 15-24 years (ASR 0.6).

**RESULTS AND CONCLUSIONS:** Incidence of childhood neuroblastoma in the North of England increased significantly over time; ASRs were 5.8 for 1968-1981 and 9.5 for 1982-1995 (rate ratio: 1.6, 95%; CI 1.2-2.3). The increase in incidence was seen in both infants and older children, and in both low stage and advanced disease. Overall 5 year survival was 15% for 1968-1981 and 40% for 1982-1995 (P < 0.0001). Significant improvements in survival were documented across different stage and age-groups, including those over 1 with stage 4 disease (0% versus 18%, P < 0.0001). Further research is needed to investigate the reasons for the increasing incidence of neuroblastoma.


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**End-of-life care for children: Bridging the gaps.**
Acute and chronic graft-versus-host disease after allogeneic peripheral- blood stem-cell and bone marrow transplantation: a meta-analysis.

Cutler C, Giri S, Jeyapalan S, Paniagua D, Viswanathan A, Antin JH.


PURPOSE: Controversy exists as to whether the incidence of graft-versus-host disease (GVHD) is increased after peripheral-blood stem-cell transplantation (PBSCT) when compared with bone marrow transplantation (BMT). We performed a meta-analysis of all trials comparing the incidence of acute and chronic GVHD after PBSCT and BMT reported as of June, 2000. Secondary analyses examined relapse rates after the two procedures. METHODS: An extensive MEDLINE search of the literature was undertaken. Primary authors were contacted for clarification and completion of missing information. A review of cited references was also undertaken. Sixteen studies (five randomized controlled trials and 11 cohort studies) were included in this analysis. Data was extracted by two pairs of reviewers and analyzed for the outcomes of interest. Meta-analyses, regression analyses, and assessments of publication bias were performed. RESULTS: Using a random effects model, the pooled relative risk (RR) for acute GVHD after PBSCT was 1.16 (95% confidence interval [CI], 1.04 to 1.28; *P* = .006) when compared with traditional BMT. The pooled RR for chronic GVHD after PBSCT was 1.53 (95% CI, 1.25 to 1.88; *P* < .001) when compared with BMT. The RR of developing clinically extensive chronic GVHD was 1.66 (95% CI, 1.35 to 2.05; *P* < .001). The excess risk of chronic GVHD was explained by differences in the T-cell dose delivered with the graft in a meta-regression model that did not reach statistical significance. There was a trend towards a decrease in the rate of relapse after PBSCT (RR = 0.81; 95% CI, 0.62 to 1.05). CONCLUSION: Both acute and chronic GVHD are more common after PBSCT than BMT, and this may be associated with lower rates of malignant relapse. The magnitude of the transfused T-cell load may explain the differences in chronic GVHD risk.

http://www.jco.org/cgi/content/full/19/16/3685
http://www.jco.org/cgi/content/abstract/19/16/3685

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DeBaun MR, Gurney JG.


Ultimately, after gathering and assessing all available evidence, pediatricians and health care policymakers must make informed decisions on whether exposure to a specific agent has the potential to cause cancer in children. In the case of DES, for which the results were clear, there was no question that the drug should be taken off the market; however, most cases of suspected carcinogens lack such clear evidence documenting cause and effect. An example of a murky topic is the suspected relationship between residential electromagnetic fields (EMFs) and childhood cancer. Epidemiologic and biologic researchers have tried for more than 2 decades to determine whether exposure to relatively high levels of EMFs poses health hazards, especially cancer in children. Although the preponderance of evidence favors a judgment that this ubiquitous environmental exposure is harmless, concerns remain in many
public circles and some scientific ones. Any proposed intervention to remove a potentially carcinogenic agent must be weighed against the cost and inconvenience to the affected community. Pediatricians are placed in a vulnerable position when faced with questions of a carcinogenic potential because of the frequency of claims in the popular literature stating that exposure to a certain product or food is associated with an increased risk for cancer in adults and possibly children. When such studies are published or, more often, released to the press, the strength of the evidence for a causal association with cancer, coupled with the context of the study, should be considered as a reasonable starting point. Better communication models of disseminating cancer-risk information are needed so that the public understands the difference between a weak study that appeared on the local news with little evidence to support a cause-and-effect relationship versus a well-designed study that was published in a peer-reviewed journal and indicates a likely cause-and-effect association.


Forgoing life-sustaining treatments: how the decision is made in French pediatric intensive care units.

Devictor DJ, Nguyen DT.


OBJECTIVES: The decision to forgo life support is frequently made in pediatric intensive care units (PICUs). A group of experts is currently preparing recommendations for guidelines concerning this decision-making process in France. We have performed a prospective study to help the experts. This study documents how children die in French PICUs and how the decision to limit life support is made. DESIGN: A multicenter, prospective, cross-sectional study. SETTING: Thirty-three multidisciplinary PICUs in university hospitals. PATIENTS: All consecutive deaths were recorded over a 4-month period. Children who died after a medical decision to forgo life-sustaining treatment were included in group 1 and children who died from other causes were included in group 2. MAIN RESULTS: A total of 264 consecutive children died, 40.1% from group 1 and 59.8% from group 2. Patients of both groups were primarily admitted for acute respiratory failure (group 1, 50.8%; group 2, 52.6%). Neurologic emergencies were more frequent in patients in group 1, whereas patients with cardiovascular failures were more frequent in group 2. When there was a question of whether to pursue life-sustaining treatment, the parents' opinions were recorded in 72.1% of cases. A specific meeting was called to make this decision in 80.1% of cases. This meeting involved the medical staff in all cases. Parents were aware of the meeting in 10.7% of cases. The conclusion of the meeting was reported to the parents in 18.7% of cases and documented in the patient's medical record in 16% of cases. Experts who were not members of the PICU staff were invited to give their opinion in 62.2% of cases. CONCLUSIONS: The decision to forgo life-sustaining treatment is frequently made for children dying in French PICUs. Guidelines must be available to help the medical staff reach this decision. Knowledge of the decision-making process in French PICUs provides the experts with information needed to elaborate such recommendations.


Factors associated with withdrawal of mechanical ventilation in a neurology/neurosurgery intensive care unit

Diringer MN, Edwards DF, Aiyagari V, Hollingsworth H.


OBJECTIVE: The objective of this study was to identify factors associated with the decision to withdraw
mechanical ventilation from patients in a neurology/neurosurgery intensive care unit. Specifically, the following factors were considered: the severity of the neurologic illness, the healthcare delivery system, and social factors. DESIGN: Retrospective analysis of prospectively collected clinical database. SETTING: Neurology/neurosurgery intensive care unit of a large academic tertiary care hospital. PATIENTS: Patients were 2,109 nonelective admissions to the neurology/neurosurgery intensive care unit who received mechanical ventilation over a period of 82 months. INTERVENTIONS: None. MEASUREMENTS AND MAIN RESULTS: The average age was 56 +/- 19.7 yrs, 53% were male, and 81% were functionally normal before admission. The median Glasgow Coma Scale score was 14, the average Acute Physiology and Chronic Health Evaluation II severity of illness score was 13.5 +/- 8.3, and probability of death was 18.2 +/- 22.0%. Mechanical ventilation was withdrawn from 284 (13.5%). Factors that were independently associated with withdrawal of mechanical ventilation were as follows: more severe neurologic injury [admission Glasgow Coma Scale score (odds ratio 0.86/point, confidence interval 0.82-0.90), diagnosis of subarachnoid hemorrhage (odds ratio 2.44, confidence interval 1.50-3.99), or ischemic stroke (odds ratio 1.72, confidence interval 1.13-2.60)], older age (odds ratio 1.04/yr, confidence interval 1.03-1.05), and higher Acute Physiology and Chronic Health Evaluation II probability of death (odds ratio 1.03/%, confidence interval 1.02-1.04). Mechanical ventilation was less likely to be withdrawn if patients were African-American (odds ratio 0.50, confidence interval 0.36-0.68) or had undergone surgery (odds ratio 0.44, confidence interval 0.2-0.67). Marital status, premorbid functional status, clinical service (neurology vs. neurosurgery), attending status (private vs. academic), and type of health insurance were not associated with decisions to withdraw mechanical ventilation. CONCLUSIONS: We conclude that decisions to withdraw mechanical ventilation in the neurology/neurosurgery intensive care unit are based primarily on the severity of the acute neurologic condition and age but not on characteristics of the healthcare delivery system. Care is less likely to be withdrawn from African-American patients or those who had surgery.


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Recent advances in cystic fibrosis.

Doull IJ.


The median life expectancy for cystic fibrosis is now over 30 years, and it is projected that in newborn infants it will become more than 40 years. The identification of the cystic fibrosis gene and its product, cystic fibrosis transmembrane conductance regulator (CFTR), has widened the spectrum of the disease from the classical case of the infant with cystic fibrosis to the elderly childless man with unexplained bronchiectasis. There is increasing evidence of the advantages of newborn screening for cystic fibrosis and subsequent specialist care. Management concentrates on optimising nutritional status and preventing lung infection and inflammation.

http://www.archdischild.com/cgi/content/full/archdischild;85/1/62
http://www.archdischild.com/cgi/content/abstract/archdischild;85/1/62

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Delayed nausea and vomiting in children receiving antineoplastics.

Dupuis LL, Lau R, Greenberg ML.


BACKGROUND: The nature and prevalence of delayed antineoplastic-induced nausea and vomiting have not been
well-described in children. This study describes the extent of delayed nausea and vomiting in children receiving antineoplastic agents as well as the drug therapies initiated in an attempt to prevent or manage it. PROCEDURE: All children receiving antineoplastics were eligible for study entry. The date and time of each emetic episode were recorded on each day antineoplastics were given and for 3 days thereafter. Nausea was self-assessed daily by children who were older than 3 years and were not developmentally delayed. Diet was also assessed daily. The emetic response, median nausea rating and median diet achieved were described. RESULTS: The emetic response of 124 children who received 174 antineoplastic cycles was evaluated. Most cycles (137/174; 79%) were not associated with delayed vomiting. Cycles which included cisplatin, carboplatin, or cyclophosphamide; involved antineoplastic therapy given over 2 or more consecutive days; or were accompanied by vomiting during the acute phase were associated with a significantly higher incidence of delayed vomiting. Moderate to severe nausea was reported on 58% (267/459) of study days. No antiemetics were given on most study days (412/522; 79%); nevertheless, most of the study days (381/412; 93%) which were unaccompanied by antiemetic support during the delayed phase were completely free from vomiting. Antiemetics were most often given as single agents (ondansetron: 54 study days; dimenhydrinate: 17 study days; dexamethasone: 6 study days). Diet was largely unaffected during the study period. CONCLUSIONS: Antineoplastic-induced delayed nausea and vomiting may be less prevalent in children than in adults. Routine antiemetic administration during the delayed phase may not be warranted in all patients. Med Pediatr Oncol 2001; 37:115-121.


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Quality-of-life measures in chronic diseases of childhood.

Eiser C, Morse R.


SCOPE OF THE REPORT: This report is concerned with the evaluation of measures broadly designed to measure quality of life (QoL) in children and adolescents, either by self-report or proxy raters. Four research questions were identified: (1) To what extent are adult measures used in the evaluation of healthcare interventions in children? (2) How appropriate are adult measures for use with children? (3) To what extent do child self-reports correspond with assessments made by parents and carers? (4) How feasible and reliable are proxy measures of QoL in different disease contexts? OBJECTIVES: (1) To review the state of the art with regard to measurement of QoL for children. (2) To make recommendations regarding the value of currently available measures for different purposes. (3) To identify further research needs. METHOD: Electronic databases were searched for the period 1980-July 1999 for articles relating to measures of QoL, health status or well-being in children (under 18 years) with chronic disease. Handsearching of relevant journals and cross-referencing with reference lists in identified articles was also carried out. Key workers in the field were contacted for additional information, and the Internet was searched for relevant websites. RESULTS: Forty-three measures were identified (19 generic and 24 disease-specific). Sixteen measures allowed for completion by children and parent/caregiver; seven only allowed for completion by a proxy, and the remainder (n = 17) allowed only for child completion. The measures were described as QoL (n = 30), health status, (n = 2), perception of illness (n = 1), life satisfaction (n = 1) and quality of well-being (n = 1). RESULTS - TO WHAT EXTENT ARE ADULT MEASURES USED IN THE EVALUATIONOF HEALTHCARE INTERVENTIONS IN CHILDREN?: Three studies were identified where adult measures were used with very few changes made for children. In 11 studies involving nine separate measures of QoL, adult measures were used as a model for work with children. RESULTS - HOW APPROPRIATE ARE ADULT MEASURES FOR USE WITH CHILDREN?: Adult measures may fail to tap the specific aspects of QoL that are important to the child. Measures based on adult work impose considerable response burden for children, in terms of length, reading skills and response scale. Wording and format of adult measures may need to be modified to account for children's cognitive and language skills. More basic research is needed to determine the level of response burden that children of different ages can manage. Assessments of difficulty (e.g. reading age) need to be routinely included with information about new measures. RESULTS - TO WHAT EXTENT DO CHILD SELF-REPORTS CORRESPOND WITH ASSESSMENTS MADE BY PARENTS AND CARERS?: Fourteen studies were identified in which concur-
dance between child and parent was investigated, often as part of the development of a new measure. There was some evidence for greater concordance between child and parent for physical functioning compared with social and emotional domains, but greater heterogeneity in the latter measures may contribute to inconsistent results. There was no simple relationship between concordance and moderating variables such as age, gender and illness, but this conclusion was addressed only very rarely. RESULTS - HOW FEASIBLE AND RELIABLE ARE PROXY MEASURES OF QOL IN DIFFERENT DISEASE CONTEXTS?: Only five papers fulfilled the review criteria. Evaluation is difficult because authors fail to justify their choice of measures, and do not report critical information such as completion rates or missing data. Use of existing measures can potentially eliminate the time and expense required to develop a comprehensive measure of QoL, but a full battery of standardised tests may be expensive in terms of time for administration and scoring. In addition, battery measures tend to be lengthy and therefore demanding for sick patients. They are not recommended for work with children. RECOMMENDATIONS FOR RESEARCH - MINIMUM CRITERIA FOR NEW MEASURES: A set of procedures needs to be established for the development of new measures. These need to draw on the experience gained in development of child and adult measures to date. Basic research to enhance understanding of how children interpret questions in QoL measures is recommended. We need to understand the differences in meaning of items between children and adults, and between children of different ages. Some attempt to develop measures for children of 6 years or more have been reported, and these should be further developed. (ABSTRACT TRUNCATED)

http://www.ncchta.org/htapubs.htm#504
http://www.ncchta.org/execsumm/summ504.htm

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A review of measures of quality of life for children with chronic illness.

Eiser C, Morse R.

Arch Dis Child 2001;84(3):205-11.

AIMS: To identify currently available generic and disease specific measures of quality of life (QoL) for work with children; and make recommendations about the future development and application of QoL measures. METHODS: Systematic searches were conducted to identify measures of QoL. Primary research papers were coded by the authors on the basis of predefined inclusion and exclusion criteria. RESULTS: Of the 137 papers included in the review, 43 involved the development of a new measure. These included 19 generic and 24 disease specific measures. Almost half the measures were developed in the USA. Measures were identified which were appropriate for children across a broad age range, and included provision for completion by different respondents (child only, parent only, or both). There were no clear distinctions between measures of QoL, health, or functional status. CONCLUSIONS: We have identified a small number of measures which fulfil basic requirements and could be used to assess QoL in clinical trials or following interventions. However, there remain a number of problems in measuring QoL in children. These include limited availability of disease specific measures; discrepancies between child and parent ratings; limited availability of measures for self completion by children; lack of precision regarding the content of domains of QoL; and the cultural appropriateness of measures developed elsewhere for children in the UK.

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Duchenne muscular dystrophy or Meryon's disease.

Emery A.
Recommendations for nonheartbeating organ donation. A position paper by the Ethics Committee, American College of Critical Care Medicine, Society of Critical Care Medicine.

Ethics Committee ACoCCMSoCCM.


Incidence of childhood acute lymphoblastic leukaemia in Yorkshire, UK.

Feltbower RG, Moorman AV, Dovey G, Kinsey SE, McKinney PA.


Between 1980 and 1998, in the north-west of England, a significant rise in childhood acute lymphoblastic leukaemia was caused by an increase in the precursor B-cell form of this disease. We analysed data on children who were diagnosed with leukaemia in Yorkshire, UK, between 1974 and 1997. The incidence of acute lymphoblastic leukaemia remained stable, although a non-significant yearly increase of 2.4% was noted for the precursor B-cell form of this disease from 1980 onwards. The precursor B-cell form accounted for 80% of all acute lymphoblastic leukaemia. Our data are not consistent with increasing incidence for precursor B-cell acute lymphoblastic leukaemia, although numbers of children with acute myeloid leukaemia are rising.

Exploring End-of-Life Care for Children With Cancer. [Editorial].

Ferguson WSMD.


Child advocacy and robust community-centered research.

Feudtner C.

Deaths attributed to pediatric complex chronic conditions: national trends and implications for supportive care services.

Feudtner C, Hays RM, Haynes G, Geyer JR, Neff JM, Koepsell TD.


Background. Children with complex chronic conditions (CCCs) might benefit from pediatric supportive care services, such as home nursing, palliative care, or hospice, especially those children whose conditions are severe enough to cause death. We do not know, however, the extent of this population or how it is changing over time. Objectives. To identify trends over the past 2 decades in the pattern of deaths attributable to pediatric CCCs, examining counts and rates of CCC-attributed deaths by cause and age (infancy: <1 year old, childhood: 1-9 years old, adolescence or young adulthood: 10-24 years old) at the time of death, and to determine the average number of children living within the last 6 months of their lives. Design/Methods. We conducted a retrospective cohort study using national death certificate data and census estimates from the National Center for Health Statistics. Participants included all people 0 to 24 years old in the United States from 1979 to 1997. CCCs comprised a broad array of International Classification of Diseases, Ninth Revision codes for cardiac, malignancy, neuromuscular, respiratory, renal, gastrointestinal, immunodeficiency, metabolic, genetic, and other congenital anomalies. Trends of counts and rates were tested using negative binomial regression. Results. Of the 1.75 million deaths that occurred in 0- to 24-year-olds from 1979 to 1997, 5% were attributed to cancer CCCs, 16% to noncancer CCCs, 43% to injuries, and 37% to all other causes of death. Overall, both counts and rates of CCC-attributed deaths have trended downward, with declines more pronounced and statistically significant for noncancer CCCs among infants and children, and for cancer CCCs among children, adolescents, and young adults. In 1997, deaths attributed to all CCCs accounted for 7242 infant deaths, 2835 childhood deaths, and 5109 adolescent deaths. Again, in 1997, the average numbers of children alive who would die because of a CCC within the ensuing 6-month period were 1097 infants, 1414 children, and 2548 adolescents or young adults. Conclusions. Population-based planning of pediatric supportive care services should use measures that best inform our need to provide care for time-limited events (perideath or bereavement care) versus care for ongoing needs (home nursing or hospice). Pediatric supportive care services will need to serve patients with a broad range of CCCs from infancy into adulthood.

Treatment demands and differential treatment of patients with cystic fibrosis and their siblings: patient, parent and sibling accounts.


Child Care Health Dev 2001;27(4):349-64.

Cystic fibrosis (CF) is a progressive disease with no known cure. Advances in diagnosis and treatment have resulted in patients living longer and thus families live with the illness for longer. Treatments are becoming increasingly demanding and are largely performed in the family home. Mothers are often reported to experience greater stress and poorer adjustment than mothers of well children or population norms. Patients and siblings are also reported to display adjustment difficulties. Siblings have rarely been included in research designs. This qualitative study investigates the impact of CF and treatment on eight patients, eight mothers, one father and eight siblings. A family systems perspective was adopted. Each individual was interviewed independently using semistructured interviews. Patients and siblings were aged between 9 and 21 years. Qualitative analyses revealed high levels of non-adherence...
(intentional and unintentional) and parental involvement in treatment, minimal involvement of siblings, and preferential treatment towards patients. Demanding treatment, coupled with the progressive nature of CF, promote high levels of parental involvement for younger children as well as older teenagers, often due to attempted or actual non-adherence. Siblings may receive less attention while patients' needs take priority. Future development of a measure of adherence suitable for children and adolescents should take into account different motivations for non-adherence, particularly the level of personal control over adherence to treatment. In addition, the potential impact of having a brother or sister with CF should not be underestimated and the needs of siblings should not go unnoticed.


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Painful peripheral neuropathy after treatment with high-dose ifosfamide.

Frisk P, Stalberg E, Stromberg B, Jakobson A.


BACKGROUND: Ifosfamide is successfully employed in the treatment of bone and soft tissue sarcomas in children and young adults. Used at high doses (HDI) the drug may cause severe multiorgan toxicity. Peripheral neuropathy is a less well-known side effect that may limit its use. We describe a 16-year-old girl with a Ewing sarcoma who was given post-operative treatment with HDI (15 mg/m2 infused over 5 days). After the second course she experienced paresthesias in both feet. After the third course she developed signs of severe toxicity in the CNS, kidneys, heart, and severe pain in her feet. PROCEDURE: Neurologic and neurophysiologic investigations, including neurographic studies of motor and sensory nerves, EMG, and thermostest, were performed in the acute phase and after 6 and 21 months, respectively. Renal and cardiac function was also assessed. RESULTS: She developed generalized weakness of the arms and legs and an extremely painful hyperesthesia of the soles. The symptoms improved gradually during follow-up but remained to some extent even after more than 2 years. Serial neurophysiologic investigations indicated classical signs of axonal neuropathy, which tended to improve during follow-up. After 18 months the glomerular filtration rate and the effective renal plasma flow were 30 and 12% of normal, respectively, while other organ functions had returned to baseline. CONCLUSIONS: Symptoms of peripheral neuropathy after HDI may herald severe multiorgan toxicity, if continued. Early administration of anesthetics through the intrathecal route should be considered in case of ifosfamide-induced painful peripheral neuropathy.


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Duration of hospitalization as a measure of cost on Children's Cancer Group acute lymphoblastic leukemia studies.


PURPOSE: We used duration of hospitalization as a surrogate for cost and event-free survival as a measure of effectiveness to estimate the cost-effectiveness ratios of various treatment regimens on Children's Cancer Group trials for acute lymphoblastic leukemia. PATIENTS AND METHODS: The analyses included 4,986 children (2 to 21 years of age) with newly diagnosed acute lymphoblastic leukemia enrolled onto risk-adjusted protocols between 1988 and 1995. Analyses were based on a model of 100 patients. The marginal cost-effectiveness ratio (hospital days per additional patient surviving event-free) was the difference in total duration of hospitalization divided by the difference in number of event-free survivors at 5 years for two regimens. Relapse-adjusted marginal cost of frontline
therapy was the difference in total duration of hospitalization for frontline therapy plus relapse therapy divided by
the difference in number of event-free survivors at 5 years on the frontline therapy for two regimens. RESULTS:
One or two delayed intensification (DI) phases, augmented therapy, and dexamethasone all improved outcome.
Marginal cost-effectiveness of these regimens compared with the control regimens was 133 days per patient for DI,
117 days per patient for double DI, and 41 days per patient for augmented therapy. Dexamethasone resulted in 17
fewer days per patient. Relapse- adjusted marginal costs were 68 days per patient for DI and 52 days for double DI.
Augmented therapy and dexamethasone-based therapy resulted in 16 and 82 fewer hospital days, respectively. The
estimated cost- effectiveness for treating any first relapse was 250 days per patient. CONCLUSION: DI, double DI,
augmented therapy, and dexamethasone-based therapy are cost-effective strategies compared with current treatment
of first relapse.

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Catheter-related thrombosis in children with cancer.

Glaser DW, Medeiros D, Rollins N, Buchanan GR.


OBJECTIVE: The prevalence of asymptomatic catheter-related thrombosis of the upper venous system in children
with cancer has not been determined. We evaluated patients with cancer and implantable central venous catheters
(ports) for this complication. STUDY DESIGN: Children with cancer undergoing port removal were eligible for this
study. Vessel patency was evaluated by contrast venography. We examined each child for physical stigmata of
thrombosis and retrospectively assessed catheter-related mechanical difficulties and infections. RESULTS: Thirty-
one ports had been placed in 24 children (aged 20 months to 18 years; median age, 9 years) with diagnoses of
leukemia/lymphoma (n = 10), solid tumor (n = 12), and histiocytosis (n = 2). Venography showed abnormalities in
12 of the 24 patients. Physical examination revealed dilated superficial veins on the chest in 3 patients. Venograms
showed abnormalities in all 3 children with prominent superficial thoracic veins. Nine of the 21 other patients had
clinically occult central venous occlusion. CONCLUSION: Fifty percent (95% CI, 30% to 70%) of children who
had implantable ports removed during or after treatment of cancer exhibited deep venous thrombosis at the site of
catheter placement. Future studies should determine the contribution of inherited and other acquired risk factors for
thrombosis and assess measures to prevent and/or treat catheter-related thrombosis in this population.

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Technology-dependent children in the community: definitions, numbers and costs.
Glendinning C, Kirk S, Guiffrida A, Lawton D.

*Child Care Health Dev 2001;27(4):321-34.*

OBJECTIVES: To assess available data on the numbers of technology-dependent children living at home in the UK and estimate the costs of caring for them. DESIGN: Data were obtained from all known secondary sources, including surveys of children with specific conditions known to specialist paediatric departments and the database of applicants to the Family Fund Trust. The costs of all services received by three hypothetical exemplar 'case study' children were calculated. RESULTS: All existing sources of data have some shortcomings. Taking these shortcomings into account, it is estimated that there may be up to 6000 technology-dependent children living at home. They appear to be disproportionately young and may be distributed unevenly between different regions of the UK. The total service costs of caring for each child will vary according to the types of technologies involved and local patterns of services, but may be as high as 150,000 pounds sterling a year. CONCLUSIONS: Because of the very high costs of services, routine information on the numbers of technology-dependent children discharged home is urgently needed. This should include details of the duration of technology dependence and the local area to which the child is discharged. New opportunities for joint service planning and purchasing should improve the co-ordination of services for these children.


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Recent advances in palliative care - Importance of palliative care for children is being increasingly recognized.

Goldman A.


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Congestive heart failure after treatment for Wilms' tumor: a report from the National Wilms' Tumor Study group.


PURPOSE: We determined the frequency of and risk factors for congestive heart failure following treatment for Wilms' tumor that included doxorubicin. PATIENTS AND METHODS: Flow sheets and medical records were reviewed to identify cases of congestive heart failure in a cohort of patients treated on National Wilms' Tumor Studies (NWTS)-1, -2, -3, and -4. The frequency of congestive heart failure was estimated using the Kaplan-Meier method. A case-control study was conducted to determine the relationship among cumulative doxorubicin dose, site(s), total dose of abdominal and thoracic irradiation, sex, and the frequency of congestive heart failure. RESULTS: The cumulative frequency of congestive heart failure was 4.4% at 20 years after diagnosis among patients treated initially with doxorubicin and 17.4% at 20 years after diagnosis among those treated with doxorubicin for their first or subsequent relapse of Wilms' tumor. The relative risk (RR) of congestive heart failure was increased in females (RR = 4.5; P = .004) and by cumulative doxorubicin dose (RR = 3.3/100 mg/m(2); P < .001), lung irradiation (RR = 1.6/10 Gy; P = .037), and left abdominal irradiation (RR = 1.8/10 Gy; P = .013). CONCLUSION: We conclude that congestive heart failure is a risk of treatment with doxorubicin for Wilms' tumor. Additional follow-up of those children treated on NWTS-4 will be necessary to determine if the decrease in dose to 150 mg/m(2) significantly reduces this risk.
Fertility preservation for children treated for cancer (1): scientific advances and research dilemmas.


Improved outcomes of children with malignancy admitted to pediatric intensive care.

Gutierrez Y, Lamelas, R., de Camargo, B.


No abstract available

The technology-dependent child.

Haffner JC, Schurman SJ.


Improvements in the provision of oxygen, mechanical ventilation, tracheostomy care, enteral and parenteral nutrition, and dialysis have expanded the population of technology-dependent children. This article attempts to review pertinent points regarding these services, including common complications. Primary care and subspecialty physicians must smooth the transition of these children to the home environment, but a comprehensive team approach is necessary for the recognition of medical complications and provision of appropriate family teaching and psychosocial supports.

Invasive procedures carried out in conscious children: contrast between North American and European paediatric oncology centres.

Hain RD, Campbell C.
AIM: To define practice in managing repeated invasive procedures in selected paediatric oncology centres in North America and Europe, especially the United Kingdom; to define and contrast concerns that shape policy making, and to contrast practice, particularly regarding procedures performed on conscious patients. METHODS: Postal survey: 118 centres of the Pediatric Oncology Group and the United Kingdom Children's Cancer Study Group received questionnaires. RESULTS: 68 questionnaires (58%) were returned (52 from North America, 12 from Europe). For all procedures, North American centres tended to use less effective techniques than European, especially for bone marrow procedures. Many North American centres reported performing these on conscious patients on at least three quarters (25%) or half (30%) the occasions. In contrast, corresponding figures for the European centres were 6% and 0%. CONCLUSIONS: Many bone marrow procedures are still carried out in the conscious patient despite the safety and effectiveness of modern anaesthetic and deep sedation techniques. There appears to be a greater reluctance to offer these to patients in North American centres than in European ones. This may reflect a misperception that the risks of adverse effects are high. Several non-pharmacological techniques are used, but they remain uncommon.

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http://www.archdischild.com/cgi/content/abstract/archdischild;85/1/12

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Effects of metabolic disorders on the brain: can these effects be reversed with bone marrow transplantation?

Hershey AD.


http://www.mosby.com/scripts/om.dll/serve?action=searchDB&searchDBfor=art&artType=fullfree&id=a116696&target=

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Attitudes and practices among pediatric oncologists regarding end-of-life care: Results of the 1998 American Society of Clinical Oncology survey.


Purpose: In 1998, the American Society of Clinical Oncology (ASCO) surveyed its membership to assess the attitudes, practices, and challenges associated with end-of-life care of patients with cancer. In this report, we summarize the responses of pediatric oncologists and the implications for care of children dying from cancer. Methods: The survey consisted of 118 questions, covering eight categories. All ASCO members in the United States, Canada, and the United Kingdom were mailed a survey, which was completed by 228 pediatric oncologists. Predictors of particular attitudes and practices were identified using stepwise logistic regression analysis. Potential predictors were age, sex, religious affiliation, importance of religious beliefs, recent death of a relative, specialty, type of practice (rural or urban, academic or nonacademic), amount of time spent in patient care, number of new patients in the past 6 months, and number of patients who died in the past year. Results: Pediatric oncologists reported a lack of formal courses in pediatric palliative care, a strikingly high reliance on trial and error in learning to care for dying children, and a need for strong role models in this area. The lack of an accessible palliative care
team or pain service was often identified as a barrier to good care. Communication difficulties exist between parents and oncologists, especially regarding the shift to end-of-life care and adequate pain control. Conclusion: Pediatric oncologists are working to integrate symptom control, psychosocial support, and palliative care into the routine care of the seriously ill child, although barriers exist that make such comprehensive care a challenge. J Clin Oncol 19:205-212. (C) 2001 by American Society of Clinical Oncology.

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The power of silence.

Himelstein BP, Jackson NL, Pegram L.


http://www.jco.org/cgi/content/full/19/19/3996

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End-of-life decision making by adolescents, parents, and healthcare providers in pediatric oncology: research to evidence-based practice guidelines.


Cancer Nurs 2001;24(2):122-34; quiz 135-6.

Participating in end-of-life decisions is life altering for adolescents with incurable cancer, their families, and their healthcare providers. However, no empirically developed and validated guidelines to assist patients, parents, and healthcare providers in making these decisions exist. The purpose of the work reported here was to use three sources (the findings of three studies on decision making in pediatric oncology, published literature, and recommendations from professional associations) to develop guidelines for end-of-life decision making in pediatric oncology. The study designs include a retrospective, descriptive design (Study 1); a prospective, descriptive design (Study 2); and a cross-sectional, descriptive design (Study 3). Settings for the pediatric oncology studies included a pediatric catastrophic illness research hospital located in the Midsouth (Studies 1 and 2); and that setting plus a children's hospital in Australia and one in Hong Kong (Study 3). Study samples included 39 guardians and 21 healthcare providers (Study 1); 52 parents, 10 adolescents, and 22 physicians (Study 2); and 43 parents (Study 3). All participants in the studies responded to six open-ended questions. A semantic content analysis technique was used to analyze all interview data. Four nurses independently coded each interview; intrarater reliability per code ranged from 68% to 100% across studies. The most frequently reported influencing factors were "information on the health and disease status of the patient," "all curative options having been attempted," "trusting the healthcare team," and "feeling support from the healthcare provider." The agreement across studies regarding influencing factors provides the basis for the research-based guidelines for end-of-life decision making in pediatric oncology. The guidelines offer assistance with end-of-life decision making in a structured manner that can be formally evaluated and individualized to meet patient and family needs.


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Parents should not be excluded from decisions to forgo life sustaining treatments!

Hoehn SK, Nelson RM.
Perinatal hospice. [Editorial].
Hoeldtke NJMDa, Calhoun BCMDb.


When the prenatal diagnosis of a lethal fetal anomaly has been established, some patients choose to continue their pregnancy. Currently, there is a paucity of medical literature addressing the specific management of families in this unique circumstance. We propose a model of care that incorporates the strengths of prenatal diagnosis, perinatal grief management, and hospice care to address the needs of these families. We discuss the identification of candidates for this form of care; the multidisciplinary team approach; and the aspects of antepartum, intrapartum, and postpartum care. Finally, we discuss some barriers that might need to be overcome when attempting to implement perinatal hospice care. (Am J Obstet Gynecol 2001;185:525-9.), (C) Mosby-Year Book Inc. 2001. All Rights Reserved.

Survivors of childhood cancer for more than twenty years.
Humpl T, Fritsche M, Bartels U, Gutjahr P.


Present health status, complications, and development of long-term survivors of childhood cancer followed for more than 20 years in a single institution were reviewed. The departmental database was searched to identify patients diagnosed with childhood cancer and consequently treated between 1965 and 1978. A total of 124 (77%) long-term survivors participated on a voluntary basis in the study. A semi-standardized interview consisted of measures evaluating the present health condition, sequelae of treatment, second malignancies, intellectual development and presence of offspring of the former patients. The majority of patients were treated with chemotherapy (82%). 67% received radiotherapy and 67% underwent surgery. A relapse of the primary tumor was diagnosed in four patients as well as a second malignancy in four other patients. In 33% of the long-term survivors one or more serious therapy-related health problems were noted. Adequate mental and intellectual development was achieved in 65%. Children treated in the early years of pediatric oncology seem to have a satisfactory outcome as viewed over the long term. Consequent ongoing follow-up is still necessary to detect health problems and enhance quality of life for subsequent generations of children with cancer.


Transdermal fentanyl for pain relief in a paediatric palliative care population.
Hunt A, Goldman A, Devine T, Phillips M, Group oboF-GS.
This multi-centre, observational study examines the efficacy of the therapeutic transdermal fentanyl system (TTS-fentanyl) in children requiring opioids for pain in life-threatening disease. Forty-one children receiving oral morphine (median 60 mg/day) transferred to transdermal fentanyl (median 25 µg/hr) in line with the manufacturer's dose conversion guidelines. Twenty-six children completed the 15-day treatment phase, seven died due to disease progression and eight were withdrawn because of adverse events, inadequate analgesia or change to parenteral opioids. At 15 days, median fentanyl dose was 75 µg/hr (range 25-250 µg/hr). No serious adverse events were attributed to fentanyl. There was a trend toward improved side effects and convenience with fentanyl. Twenty-three of 26 parents (three missing) and 25 of 26 investigators considered transdermal fentanyl to be better than previous treatment. For all records available (at 15 days or on withdrawal if earlier), 75% (27/36) reported that fentanyl treatment was "good" or "very good". The findings suggest that transdermal fentanyl is both effective and acceptable to children and families.

Paediatric palliative care: distinctive needs and emerging issues.

Hynson JL, Sawyer SM.


Palliative medicine has developed as a specialized field of practice in recent decades but the focus has been very much on older adults with incurable malignancies. The needs of dying children have not been addressed adequately and the question of who is best placed to provide care to this group remains the subject of some contention. Although the principles of palliative care apply equally to children, a number of fundamental differences influence their application in the paediatric setting. These include a heterogeneous patient population, physiological factors, developmental issues, parental involvement in care giving and decision making and the desire of most paediatricians to maintain close involvement with their patients. Families generally prefer home care and even quite severe symptoms can be managed in this environment with appropriate planning, expertise and support. The delivery of effective palliative care in the paediatric setting is contingent upon overcoming barriers between hospital and community and sharing expertise between paediatricians and palliative care physicians. Research is also required to increase the evidence base for practice.

Hyperbaric oxygenation for cerebral palsy.

James PB.


Metastatic Ewing sarcoma/PNET of bone at diagnosis: prognostic factors-- a report from Saudi Arabia.

BACKGROUND: To evaluate outcome and prognostic factors in Saudi Arabian patients with metastatic Ewing sarcoma and PNET of bone (PMES) at diagnosis. PROCEDURE: Ninety-nine of 304 (33%) consecutive patients with Ewing sarcoma and PNET of bone registered at our centre from 1975 to 1998, had metastatic disease at registration and 93 were available for analysis. The maximum x-axis diameter of the primary tumor was used as the measure of primary tumor size. Usually a trial of systemic treatment was undertaken before a decision was made on local treatment. Standard chemotherapy regimens were used in all treated patients. Forty-five (49%) patients did not receive radical local treatment due to an inadequate response to chemotherapy, or a decision not to undergo more palliative treatment. Radical treatment of the primary site was radiation alone 41 (79\%), resection alone 7 (13\%), and resection and radiation 4 (8\%). RESULTS: The 5-year survival rates were 9\% for all 93 evaluable patients, 16\% for 52 patients who received chemotherapy and radical local treatment, 0\% for 41 patients who received lesser treatment, 19\% for 43 patients with lung metastases alone, and 0\% (P = 0.002) for 50 patients with other sites involved. For 60 patients with imaging data, 5-year survivals were 34 and 0\% when the maximum transverse diameter of the primary tumor was < 10 cm (N = 20) and ≥ 10 cm (N = 40), respectively. CONCLUSIONS: Small primary tumor size and the presence of lung metastases alone were the only significant favorable prognostic factors. Earlier diagnosis will be the basis for better results.


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Characterisation of breathing and associated central autonomic dysfunction in the Rett disorder.


AIM: To investigate breathing rhythm and brain stem autonomic control in patients with Rett disorder. SETTING: Two university teaching hospitals in the United Kingdom and the Rett Centre, Sweden. PATIENTS: 56 female patients with Rett disorder, aged 2-35 years; 11 controls aged 5-28 years. DESIGN: One hour recordings of breathing movement, blood pressure, ECG R-R interval, heart rate, transcutaneous blood gases, cardiac vagal tone, and cardiac sensitivity to baroreflex measured on-line with synchronous EEG and video. Breathing rhythms were analysed in 47 cases. RESULTS: Respiratory rhythm was normal during sleep and abnormal in the waking state. Forced and apneustic breathing were prominent among 5-10 year olds, and Valsalva breathing in the over 18 year olds, who were also most likely to breathe normally. Inadequate breathing peaked among 10-18 year olds. Inadequate and exaggerated breathing was associated with vacant spells. Resting cardiac vagal tone and cardiac sensitivity to baroreflex were reduced. CONCLUSIONS: Labile respiratory rhythms and poor integrative inhibition in Rett disorder suggest brain immaturity. Linking this to an early monoaminergic defect suggests possible targets for the MECP2 gene in clinical intervention. Exaggerated and inadequate autonomic responses may contribute to sudden death.

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Alleviating the suffering of seriously ill children.

Kane JR, Primomo M.
Modern medicine has largely focused on the physical aspects of disease, aggressively attacking the illness, often at the expense of caring for pain and suffering. Medical interventions based solely on the diagnosis and treatment of disease limit the medical care of the severely ill child. Such an approach is particularly detrimental when caring for the terminally ill. Successful care of children with chronic, life-threatening, or terminal illnesses requires a comprehensive assessment of their physical, psychological, and spiritual needs as well as a process of collaboration between members of the multiple disciplines involved in the care of the patient and the family unit as a whole. Supportive/palliative care serves as a bridge between a scientific (disease-oriented) and humanistic (person-oriented) approach to patient care. Bridging this gap early in the course of life-threatening illness is essential for successful palliative intervention to relieve suffering and improve the quality of life for the child and his or her family. A model that introduces supportive, palliative, and hospice services into the mainstream of medical therapy is emphasized as a standard for the care of all children with significant chronic, life-threatening, or terminal illness. This article expands on a previous paper published in the American Journal of Hospice & Palliative Care (Kane JR, Barber RG, Jordan M, et al.: Supportive/palliative care of children suffering from life-threatening and terminal illness. May/June 2000; 17(3): 165-172).


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Complementary and alternative medicine for children: does it work?

Kemper KJ.


http://www.ewjm.com/cgi/content/full/174/4/272

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Negotiating lay and professional roles in the care of children with complex health care needs.

Kirk S.


Negotiating lay and professional roles in the care of children with complex health care needs Background. Children with complex health care needs are now being cared for at home as a result of medical advances and government policies emphasizing community-based care. The parents of these children are involved in providing care of a highly technical and intensive nature that in the past would have been the domain of professionals (particularly nurses). Aims of the paper. To assess how the transfer of responsibility from professionals to parents was negotiated, the tensions and contradictions that can ensue, and the implications for professional nursing roles and relationships with parents. Design. Using a Grounded Theory methodology, in-depth interviews were conducted with 23 mothers, 10 fathers and 44 professionals to gain insight into the experience of caring for children and supporting families in the community. Findings. From the parents' perspective, their initial assumption of responsibility for the care of their child was not subject to negotiation with professionals. Prior to discharge, parents' feelings of obligations, their strong desire for their child to come home, and the absence of alternatives to parental care in the community, were the key motivating factors in their acceptance of responsibility for care-giving from professionals. The professionals
participating in the study had concerns over whether this group of parents was given a choice in accepting responsibility and the degree of choice they could exercise in the face of professional power. However, it was following the initial discharge, as parents gained experience in caring for their child and in interacting with professionals, that role negotiation appeared to occur. Conclusion. This study supports other research that has found that professionals’ expectations of parental involvement in the care of sick children role can act as a barrier to negotiation of roles. In this study, parental choice was also constrained initially by parents' feelings of obligation and by the lack of community services. Nurses are ideally placed to play the central role not only in ensuring that role negotiation and discussion actually occurs in practice, but also by asserting the need for appropriate community support services for families. Being on home territory, and in possession of expertise in care-giving and in managing encounters with professionals, provided parents with a sense of control with which to enter negotiations with professionals. It is important that changes in the balance of power does not lead to the development of parent-professional relationships that are characterized by conflict rather than partnership.


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Parental wishes and poor outcomes in preterm infants.

Koh TH.


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Does palliative care palliate?

Kornblith AB.


http://www.jco.org/cgi/content/full/19/8/2111

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Decision making in extreme situations involving children: withholding or withdrawal of life supporting treatment in paediatric care. Statement of the ethics working group of the Confederation of the European Specialists of Paediatrics (CESP).

Kurz R.


Paediatricians increasingly find themselves in situations in which decisions must be made regarding withholding or withdrawing life-supporting treatment in the care of a paediatric patient. There comes a point when the artificial prolongation of life only contributes to extending the act of dying and with it the suffering of the patient,
parents/guardians, family, and caregivers. Life is the most precious gift. Every person, every child, has the right to live. Similarly, every person, every child, has the right to die with dignity. The paediatrician has an obligation to each patient to preserve life, promote health, and treat illness. In recent years the capacity of medicine to prolong life has increased exponentially. In situations that are extremely burdensome, and where there is no chance for effective therapy, the paediatrician has an obligation to protect the dignity of the patient in the act of dying.

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Mothers' grief following the death of a child.

Laakso H, Paunonen-Ilmonen M.


Aim of the study. Research has shown that caring for a dying child is among the hardest and more demanding tasks in nursing, because the staff are forced to manage their heavy work with inadequate skills and experience. This article deals with the findings of a recent study, the purpose of which was to analyse the mother's grief and coping with grief following the death of a child under the age of 7 years. Design. Data were collected from mothers using a survey (n=91) and an interview (n=50). As the topic was very sensitive ethically and emotionally, survey data were collected first and the mothers were asked to give their consent to taking part in an interview. The study employed both quantitative and qualitative methods. The data were analysed using statistical methods and content analysis. However, only the qualitative part of the study is presented in this article. Findings. The findings show that nursing staff had skills to support grieving mothers, but that there were many feelings and experiences of grief that remained unidentified by staff. The staff's ability to meet the mothers' individual needs while the child was in hospital and after the child's death was inadequate. The information received from staff was perceived to be insufficient or offensive to mothers. Conclusions. The development of basic and further education and of various support measures would enable the staff to better cope with their work. Focusing on interactive skills and meeting the patient's individual needs using reflective practice would improve the quality of care. Communication and collaboration between different occupational groups should be promoted, because mothers were dissatisfied with dissemination of information, and ambiguous responsibilities between different occupational groups hampered the acquisition of information.

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Cognitive functions of adolescent childhood cancer survivors assessed by event-related potentials.

Lahteenmaki PM, Holopainen I, Krause CM, Helenius H, Salmi TT, Heikki LA.


BACKGROUND: Neurophysiological methods were applied to examine subtle central nervous system (CNS) adverse effects for adolescent childhood cancer survivors. We analyzed auditory event-related potentials (ERPs)-P300 and MMN/P3a complex-to find out whether there was impaired attention orientation in asymptomatic cancer survivors, and whether these ERP methods could be used as more objective tools in detecting those survivors who might need academic testing. Previous clinical studies of P300 have focused on leukemia survivors. MMN for cancer survivors has not been reported. PROCEDURE: The subjects were survivors of childhood leukemia (n=11) and solid tumors (n=8), as well as healthy controls (n=10). The mean age was 15.5 years for survivors and 15.9 years for controls. Pure sine-wave tones (500 and 553 Hz, 100 ms) were used as stimuli in an oddball paradigm. The
ERPs to frequency change were measured. MMN recordings were performed in a passive non-attended situation where the subject was watching a voiceless video cartoon. P300 was produced thereafter, but in an active attend situation, by the same auditory oddball paradigm as MMN. RESULTS: A significant difference was detected between the groups for the latency of P300 at electrodes Cz (P = 0.03) and C4 (P = 0.05). The cancer survivors had prolonged P300 latencies as an indication of prolonged short-term memory processing. The area and latency parameters of MMN did not differ significantly between the study groups, but in cancer survivors, the area and the mean amplitude of the subsequent P3a wave were diminished. The results indicate that the discrimination process was not as easy for the survivors as for the controls. However, it seems that in cancer survivors the basic mechanism starting attention shift to novel stimuli is not impaired. CONCLUSIONS: These results indicate that it is important to carefully evaluate the proper methods for the teaching of children who are survivors of malignancies. The auditory information may not always lead to the best possible learning results.


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Does salvage therapy influence outcome?

Langer CJ.


http://www.jco.org/cgi/content/full/19/7/2108

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Prevalence and morbidity associated with non-malignant, life-threatening conditions in childhood.

Lenton S, Stallard P, Lewis M, Mastroyannopoulou K.

Child Care Health Dev 2001;27(5):389-98.

OBJECTIVE: To determine the prevalence of non-malignant life-threatening illness in childhood and associated morbidity in the affected child and their family members. DESIGN: Cross-sectional survey. SETTING: Bath Clinical Area (total population 411 800). SUBJECTS: Children aged 0-19 years. RESULTS: One hundred and twenty-three children were identified, giving a prevalence of 1.2/1000 children. Morbidity assessed in 93 children showed 60% in pain or discomfort, 35% unable to walk and 25% with severe cognitive impairment. Mental health problems were found in 54% of mothers and 30% of fathers, and significant emotional and behavioural problems in 24% of healthy siblings. CONCLUSIONS: Non-malignant life-threatening illness is more prevalent than reported in previous studies. Considerable morbidity is experienced by the child and their family. An individual and family approach is required. KEY MESSAGES (1) THE PREVALENCE OF NON-MALIGNANT LIFE-THREATENING ILLNESS IS FOUR TIMES GREATER THAN PREVIOUS ESTIMATES. (2) THIS GROUP OF CONDITIONS HAVE SIGNIFICANT IMPLICATIONS FOR ALL FAMILY MEMBERS. (3) EARLY COMPREHENSIVE ASSESSMENT AND ACCESS TO EFFECTIVE INTERVENTIONS MAY PRE-EMPT LATER PROBLEMS:


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Friedreich's ataxia presenting after cardiac transplantation.

Leonard H, Forsyth R.


A 4 year old boy underwent cardiac transplantation because of cardiomyopathy with ischaemia. Following transplantation he developed neurological signs of Friedreich's ataxia and the diagnosis was confirmed with genetic testing. Cardiomyopathy is a rare presentation of Friedreich's ataxia and to our knowledge this is the first reported transplant operation for the cardiomyopathy associated with this condition.

http://www.archdischild.com/cgi/content/full/archdischild;84/2/167
http://www.archdischild.com/cgi/content/abstract/archdischild;84/2/167

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Experience with neonatal palliative care consultation at the Medical College of Wisconsin-Children's Hospital of Wisconsin.

Leuthner SR, Pierucci R.


At Children's Hospital of Wisconsin there is a pediatric palliative care consultation service that serves a diverse patient population, including infants. However, the value of a palliative care consultation for infants has not been well evaluated. We performed a retrospective, case series, descriptive chart review of infants in our neonatal intensive care unit (NICU) who received palliative care consults between January 1996 and June 1998. We specifically looked at their diagnoses, the timing of consults, reasons that consultations were ordered, what recommendations were made, and the subsequent outcomes. During the series period there were 898 admissions to the NICU, 51 neonatal deaths, and 12 neonatal palliative care consultations. The diagnostic categories for those with a palliative care referral included prematurity, lethal anomalies, and catastrophic or overwhelming illness. Reasons for the consultations were organization of home hospice, facilitation of medical options, such as do-not-resuscitate (DNR) orders and treatment withdrawal, facilitation of comfort measures, and grief/loss issues. Recommendations that the palliative care staff made fell into four categories: advance directive planning, the optimal environment for supporting neonatal death, comfort and medical care, and psychosocial support. This series is a description of what a palliative care service can offer for terminally ill infants in an NICU. We speculate that such consults can more consistently and comprehensively provide appropriate end-of-life care for these patients and their families.


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Immunization of HIV-infected children with varicella vaccine.


OBJECTIVE: To determine the safety and immunogenicity of varicella vaccine in children with human immunodeficiency virus (HIV) infection. Children (n = 41) who were mildly affected by HIV (Centers for Disease Control and Prevention stage N1 or A1) and had no history or serum antibody indicative of prior varicella infection...
were immunized with two doses of live attenuated varicella vaccine. RESULTS: A minority of the vaccine recipients had mild local or systemic reactions. Vaccination had no effect on the clinical stage of HIV or the HIV RNA plasma load. CD4 cell percentage and CD4 cell count were marginally decreased at week 4 after the first vaccination; this effect was no longer present at week 8 after vaccination. Two months after the second dose of vaccine, 60% of vaccine recipients had anti-varicella antibody in their serum, and 83% had a positive lymphocyte proliferation assay response to varicella antigen. CONCLUSION: On the basis of its safety and immunogenicity, varicella vaccine should be considered in the childhood vaccines given to mildly affected HIV-infected children.

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Paying attention to death.

Levy MM.


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Normal pulse oximeter reading in a cyanotic infant.


A newborn infant who presented with central cyanosis was found to have hereditary methaemoglobinemia. The pulse oximeter readings and physical findings were incompatible. Clinical assessment remains an important part in the management of such cases.


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Health status of children with moderate to severe cerebral palsy.


The aim of the study was to evaluate the health of children with cerebral palsy (CP) using a global assessment of quality of life, condition-specific measures, and assessments of health care use. A multicenter population-based cross-sectional survey of 235 children, aged 2 to 18 years, with moderate to severe impairment, was carried out using Gross Motor Function Classification System (GMFCS) levels III (n = 56), IV (n = 55), and V (n = 122), This study group scored significantly below the mean on the Child Health Questionnaire (CHQ) for Pain, General Health, Physical Functioning, and Impact on Parents. These children used more medications than children without CP from a national sample. Fifty-nine children used feeding tubes, Children in GMFCS level V who used a feeding tube had the lowest estimate of mental age, required the most health care resources, used the most medications, had the most
respiratory problems, and had the lowest Global Health scores, Children with the most severe motor disability who have feeding tubes are an especially frail group who require numerous health-related resources and treatments. Also, there is a relationship among measures of health status such as the CHQ, functional abilities, use of resources, and mental age, but each appears to measure different aspects of health and well-being and should be used in combination to reflect children's overall health status.

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**Pediatric HIV disease.**

listed Na.


No abstract available

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**Lumbar puncture in pediatric oncology: Conscious sedation vs. general anesthesia.**

Ljungman G, Gordh T, Sorensen S, Kreuger A.

*Medical and Pediatric Oncology* 2001;36(3):372-379.

Background. Lumbar punctures (LP) generally have been performed with conscious sedation (CS) but are increasingly performed using general anesthesia (GA) owing to the belief that this is less distressing. The aim of this study was to compare these two methods concerning distress, discomfort, pain, well-being and security after the LP, and problems with the LP. Procedure. Twenty-five children with cancer participated in this prospective, randomized, crossover study. Children, parents, and nurses completed a visual analogue scale questionnaire to evaluate the efficacy of CS and GA. In addition, sedation and anesthesia protocols were reviewed. Results. The two methods seemed to be equivalent concerning distress, discomfort, pain, well-being and security after the procedure, and procedure problems. Most children (80%), parents (66%), and nurses (58%) preferred LP in CS. However, the LP was not performed in CS in five cases because the child did not cooperate. Younger children less often preferred CS. Conclusions. Outcomes for CS and GA in LP were similar. Although there were failures with the CS model, most preferred it to GA. LP in CS also saved time and medical resources. An alternative approach would be to have a pediatric anesthesiologist available at the oncology ward for these elective LPs to provide the sedation or anesthesia required by each individual. Med. Pediatr. Oncol. 36:372-379, 2001. (C) 2001 Wiley-Liss, Inc.

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**The role of nurses in the management of children with epidermolysis bullosa.**


The dystrophic form of congenital epidermolysis bullosa causes skin lesions resembling those of patients with severe burns. The loosening of the skin is permanent and occurs repeatedly, in a series of flare-ups. The mucous membranes are particularly severely affected. Skin care requires numerous personnel and pain management is of prime importance. The management of this condition is therefore highly specialised. The prognosis is very poor with dystrophic epidermolysis bullosa typically resulting in the death of the patient. (C) 2001 Editions scientifiques et medicales Elsevier SAS.

Care of the potential pediatric organ donor.

Lutz-Dettinger N, de Jaeger A, Kerremans I.


Although all of this information may create the impression that caring for a potential organ donor is an exceedingly complex task, in the authors’ experience, this often is not true, and much energy can--and should--be devoted to the care of the bereaved family. Of crucial importance are the early recognition of brain death and the consequent radical switch of the treatment goal from preservation of the patient’s brain and life to preservation of organs for the lives of others. Care for the donor is the natural extension of care for a critically ill or injured patient. During the foregoing discussion, the authors had to stress the absence of sound evidence on many points. Because many reports originate from transplant centers dedicated to a specific organ, gaining a comprehensive view on management options in the ICU further is hampered. Thus, this situation leaves another field in which investigations originating from pediatric intensivists could provide evidence urgently needed to make optimal choices. The next decade should see the thyroid hormone controversy solved by at least one controlled prospective study and the differential applicability of inotropic, vasoactive, or fluid-centered strategies. It seems self-evident that only graft survival and related parameters can form adequate endpoints for future studies.

Influenza vaccination in HIV-infected children.

Marczynska M, Brydak LB, Machala M, Oldakowska A, Zegadlo M.


Who can decide what is in a child's best interest? A problem-solving approach.

Masera G.

Clinical research in palliative care: patient populations, symptoms, interventions and endpoints.

Mazzocato C, Sweeney C, Bruera E.


Clinical trials in palliative care involve multiple issues relating to patient populations, interventions and endpoints. Careful data collection and analysis of variables are vital for good clinical research in this complex area.


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Death in the family: Adapting a family systems framework to the grief process.

McBride J, Simms S.

_American Journal of Family Therapy 2001;29(1):59-73._

This paper presents a framework for adapting a family systems approach to the grief process. The framework places the bereaved and therapist in the larger context. The therapist builds a relationship around each family member's unique experience with death, and uses the grief timeline to facilitate timely systemic interventions. This perspective helps the therapist navigate pitfalls and remain strategically positioned to address grief sensitivity and effectively in psychotherapy.

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Trained volunteers for families coping with a child with a life-limiting condition.

McGrath P.


One hundred and seven families with a child diagnosed with a life-limiting condition, from all over Queensland (Australia), were surveyed on their need for a trained volunteer. Their comments provide important insights into an area which has not previously been researched or documented. In summary, their responses indicate that many families have only minimal or no support, and are coping with extraordinary physical, emotional and social demands from the child's condition and treatment. The majority of the participants are very positive about the need for a trained volunteer and are clear about the activities that trained volunteers could provide assistance with. These activities range from practical assistance such as baby-sitting and help with household chores and errands to emotional support. Not all families would be comfortable using a volunteer, and some respondents did outline perceived obstacles to including a volunteer in family activities. For others there were comments about why volunteers would be perceived as unproblematic. Respondents specified important criteria that would need to be addressed in volunteer training. The largest number of respondents were coping with cystic fibrosis and muscular dystrophy. It is the hope and expectation that, with the communication of the insights gathered from this geographically diverse group of families, encouragement will be given to others working in this area to explore the possibility of establishing outreach volunteer programmes for such families.
Lingering death after treatment withdrawal in the neonatal intensive care unit. [Article].

McHaffie HE, Lyon AJ, Fowlie PW.


**OBJECTIVE:** To explore parents' perceptions of treatment withdrawal and the dying process., **DESIGN:** Face to face interviews with 59 sets of parents of 62 babies in the East of Scotland three months and 13 months after death., **RESULTS:** 22% of the parents expressed reservations about the length of the dying process, which they reported in these instances had taken from three to 36 hours. Deaths that medical teams had predicted would be quick had, according to the parents' recollections, taken from 1.5 to 31 hours. When a baby died swiftly, this seemed to confirm the wisdom of the decision to stop. When babies lingered, doubts were raised., **CONCLUSIONS:** Parents need to be adequately prepared for what may happen after treatment withdrawal. The debate should be reopened about the best way to manage protracted deaths in line with parental need., Copyright (C) 2001 by Archives of Disease in Childhood.

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Long-term comparative trial of positive expiratory pressure versus oscillating positive expiratory pressure (flutter) physiotherapy in the treatment of cystic fibrosis.

McIlwaine PM, Wong LT, Peacock D, Davidson AG.


**OBJECTIVE:** The objective was to evaluate the long-term effects of physiotherapy with an oscillating positive pressure device ("flutter") compared with physiotherapy with the use of a positive expiratory pressure (PEP) mask in patients with cystic fibrosis (CF). **Study design:** Forty children with CF were randomly assigned to performing physiotherapy with the PEP mask or the flutter device for 1 year. Clinical status, pulmonary function, and compliance were measured at regular intervals throughout the study. **RESULTS:** The flutter group demonstrated a greater mean annual rate of decline in forced vital capacity compared with the PEP group (-8.62 +/- 15.5 vs 0.06 +/- 7.9; P =.05) with a similar trend in forced expiratory volume in 1 second (- 10.95 +/- 19.96 vs -1.24 +/- 9.9; P =.08). There was a significant decline in Huang scores (P =.05), increased hospitalizations (18 vs 5; P =.03), and antibiotic use in the flutter group. **CONCLUSION:** Flutter was not as effective in maintaining pulmonary function in this group of patients with CF compared with PEP and was more costly because of the increased number of hospitalizations and antibiotic use.

http://www.mosby.com/scripts/om.dll/serve?action=searchDB&searchDBfor=art&artType=abs&id=a114017&target=

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Screening for Wilms tumor and hepatoblastoma in children with Beckwith- Wiedemann syndromes: a cost-effective model.


BACKGROUND: We undertook a cost-benefit analysis of screening for Wilms tumor and hepatoblastoma in children with Beckwith-Wiedemann syndrome (BWS), a known cancer predisposition syndrome. The purpose of this analysis was twofold: first, to assess whether screening in children with BWS has the potential to be cost-effective; second, if screening appears to be cost-effective, to determine which parameters would be most important to assess if a screening trial were initiated. PROCEDURES: We used data from the BWS registry at the National Cancer Institute, the National Wilms Tumor Study (NWTS), and large published series to model events for two hypothetical cohorts of 1,000 infants born with BWS. One hypothetical cohort was screened for cancer until a predetermined age, representing the base case. The other cohort was unscreened. For our base case, we assumed: (a) sonography examinations three times yearly (triannually) from birth until 7 years of age; (b) screening would result in one stage shift downward at diagnosis for Wilms tumor and hepatoblastoma; (c) 100% sensitivity and 95% specificity for detecting clinical stage I Wilms tumor and hepatoblastoma; (d) a 3% discount rate; (e) a false positive result cost of $402. We estimated mortality rates based on published Wilms tumor and hepatoblastoma stage specific survival. RESULTS: Using the base case, screening a child with BWS from birth until 4 years of age results in a cost per life year saved of $9,642 while continuing until 7 years of age results in a cost per life-year saved of $14,740. When variables such as cost of screening examination, discount rate, and effectiveness of screening were varied based on high and low estimates, the incremental cost per life-year saved for screening up until age four remained comparable to acceptable population based cancer screening ranges (< $50,000 per life year saved). CONCLUSIONS: Under our model's assumptions, abdominal sonography examinations in children with BWS represent a reasonable strategy for a cancer screening program. A cancer screening trial is warranted to determine if, when, and how often children with BWS should be screened and to determine cost-effectiveness in clinical practice.


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Late mortality experience in five-year survivors of childhood and adolescent cancer: the Childhood Cancer Survivor Study.


PURPOSE: Survivors of childhood and adolescent cancer are at risk for long-term effects of disease and treatment. The Childhood Cancer Survivor Study assessed overall and cause-specific mortality in a retrospective cohort of 20,227 5-year survivors. PATIENTS AND METHODS: Eligible subjects were individuals diagnosed with cancer (from 1970 to 1986) before the age of 21 who had survived 5 years from diagnosis. Underlying cause of death was obtained from death certificates and other sources and coded and categorized as recurrent disease, sequelae of cancer treatment, or non-cancer-related. Age and sex standardized mortality ratios (SMRs) were calculated using United States population mortality data. RESULTS: The cohort, including 208,947 person-years of follow-up, demonstrated a 10.8-fold excess in overall mortality (95% confidence interval, 10.3 to 11.3). Risk of death was statistically significantly higher in females (SMR = 18.2), individuals diagnosed with cancer before the age of 5 years (SMR = 14.0), and those with an initial diagnosis of leukemia (SMR = 15.5) or CNS tumor (SMR = 15.7). Recurrence of the original cancer was the leading cause of death among 5-year survivors, accounting for 67% of deaths. Statistically significant excess mortality rates were seen due to subsequent malignancies (SMR = 19.4), along with cardiac (SMR = 8.2), pulmonary (SMR = 9.2), and other causes (SMR = 3.3). Treatment-related associations were present for subsequent cancer mortality (radiation, alkylating agents, epipodophyllotoxins), cardiac mortality (chest irradiation, bleomycin), and other deaths (radiation, anthracyclines). No excess mortality was observed for external causes (SMR = 0.8). CONCLUSION: While recurrent disease remains a major contributor to late mortality in 5-year survivors of childhood cancer, significant excesses in mortality risk associated with treatment-related complications exist up to 25 years after the initial cancer diagnosis.

http://www.jco.org/cgi/content/full/19/13/3163
Survival after first esophageal variceal hemorrhage in patients with biliary atresia.

Miga D, Sokol RJ, Mackenzie T, Narkewicz MR, Smith D, Karrer FM.


**OBJECTIVE:** To determine the influence of the new onset of esophageal variceal hemorrhage (EVH) on transplant-free survival in children with biliary atresia and to examine variables that predicted survival after the onset of EVH.

**METHODS:** Retrospective chart review of 134 patients with biliary atresia who underwent portoenterostomy between 1973 and 1992 at a single institution; 29% had EVH.

**RESULTS:** The risk of death or need for liver transplantation was 50% at 6 years after the initial episode of EVH. Patients with a serum bilirubin concentration < or =4 mg/dL at the first episode of EVH had transplant-free survival of >80% for 4 years after this episode, those with bilirubin levels >4 to 10 mg/dL had 50% survival at 1 year, and those with bilirubin levels >10 mg/dL had 50% survival at 4 months. The risk of death or transplant for a child with EVH and total serum bilirubin levels >10 mg/dL was 12.0 (95% CI: 6.0, 24.1), 4 to 10 mg/dL was 7.2 (3.1, 16.7), and < or =4 mg/dL was 0.6 (0.1, 3.1) times the risk of a same-aged child who did not have EVH.

**CONCLUSIONS:** Children with biliary atresia and first EVH episode have a variable prognosis related to total serum bilirubin concentration at the time of the episode.

Simultaneous monitoring of gastric and oesophageal pH reveals limitations of conventional oesophageal pH monitoring in milk fed infants.

Mitchell DJ, McClure BG, Tubman TR.


**BACKGROUND:** Monitoring oesophageal pH conventionally detects "acid reflux" (pH less than 4). The pH of the gastric contents determines whether or not reflux can be detected. **AIM:** To monitor gastric and oesophageal pH simultaneously in order to determine the effect of milk feeds on gastric pH and how this would influence interpretation of the oesophageal pH record. **METHODS:** Milk fed infants for whom oesophageal pH monitoring was requested underwent simultaneous gastric and oesophageal pH monitoring using a dual channel pH probe. **RESULTS:** Twenty of 24 records were technically satisfactory. Mean reflux index was 1.0%, range 0.0-4.0%. Gastric pH was less than 4 for 24.5% (range 0.6-69.1%) of the total time. The average time the gastric pH was greater than 4 after feeds was 130 minutes (range 29-212 minutes). The corrected reflux index (limited to the time the gastric pH was less than 4) was 2.6% (range 0.0-11.0%). **CONCLUSION:** The pH of the gastric contents may be greater than 4 for prolonged intervals, during which oesophageal pH monitoring using current criteria cannot detect reflux nor correlate it with clinical events. A low reflux index may reflect prolonged buffering of gastric acidity rather than the absence of reflux.
Paediatric brain death in Japan.

Miyasaka K, Takeuchi K, Takeshita H.


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Decreasing late mortality among five-year survivors of cancer in childhood and adolescence: a population-based study in the Nordic countries.


PURPOSE: To assess the risk of death in patients who survive more than 5 years after diagnosis of childhood cancer and to evaluate causes of death in fatal cases. PATIENTS AND METHODS: This was a population-based study in the five Nordic countries (Denmark, Finland, Iceland, Norway, and Sweden) using data of the nationwide cancer registries and the cause-of-death registries. The study cohort included 13,711 patients who were diagnosed with cancer before the age of 20 years between 1960 and 1989 and who survived at least 5 years from diagnosis. By December 31, 1995, 1,422 patients had died, and death certificates were assessed in 1,402. Standardized mortality ratios (SMRs) for validated causes of death were calculated based on 156,046 patient-years at risk. RESULTS: The overall SMR was 10.8 (95% confidence interval [CI], 10.3 to 11.5), mainly due to high excess mortality from the primary cancer. SMR for second cancer was 4.9 (95% CI, 3.9 to 5.9) and was 3.1 (95% CI, 2.8 to 3.5) for noncancer death. The pattern of causes of death varied markedly between different groups of primary cancer diagnoses and was highly dependent on time passed since diagnosis. Overall late mortality was significantly lower in patients treated during the most recent period of time, 1980 to 1989, compared with those treated from 1960 to 1979 (hazard ratio, 0.61; 95% CI, 0.54 to 0.70), and there was no increase in rates of death due to cancer treatment. CONCLUSION: Long-term survivors of childhood cancer had an increased mortality rate, mainly dying from primary cancers. However, modern treatments have reduced late cancer mortality without increasing the rate of therapy-related deaths.

http://www.jco.org/cgi/content/full/19/13/3173
http://www.jco.org/cgi/content/abstract/19/13/3173

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Risks of young age for selected neurocognitive deficits in medulloblastoma are associated with white matter loss.


PURPOSE: To test the hypothesis that inadequate development of normal-appearing white matter (NAWM) is associated with the relationship between young age at the time of craniospinal irradiation (CRT) and deficient neurocognitive performance in survivors of childhood medulloblastoma. PATIENTS AND METHODS: Forty-two patients treated since 1985 participated in this cross-sectional study. All had been treated with CRT with or without chemotherapy and had survived 1 or more years after treatment. Neurocognitive evaluations were conducted with
tests of intellect (intelligent quotient; IQ), verbal memory, and sustained attention. Quantitative magnetic resonance imaging, using a hybrid neural network, assessed the volume of NAWM. RESULTS: Neurocognitive test results were below normal expectations for age at the time of testing. A young age at CRT was significantly associated with worse performance on all neurocognitive tests except that of verbal memory. An increased time from completion of CRT was significantly associated with worse performance on all neurocognitive tests except that of sustained attention. After statistically controlling for the effects of time from CRT, we examined the association of NAWM with neurocognitive test results. These analyses revealed that NAWM accounted for a significant amount of the association between age at CRT and IQ, factual knowledge, and verbal and nonverbal thinking, but not sustained attention or verbal memory. CONCLUSION: The present results suggest that, at least for some cognitive functions, deficient development and/or loss of NAWM after CRT may provide a neuroanatomical substrate for the adverse impact of a young age at the time of CRT.

http://www.jco.org/cgi/content/full/19/2/472
http://www.jco.org/cgi/content/abstract/19/2/472

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Musculoskeletal manifestations of neuroblastoma at diagnosis.

Murthi GV, Azmy A, Carachi R, Goel K.


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Hyperbaric oxygenation for cerebral palsy.

Neubauer RA.

Lancet 2001;357(9273):2052; discussion 2053.


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Insulin improves clinical status of patients with cystic-fibrosis-related diabetes mellitus.

Nousia-Arvanitakis S, Galli-Tsinopoulou A, Karamouzis M.


Cystic-fibrosis-related diabetes mellitus is frequently underdiagnosed and associated with deterioration of overall clinical status. The purpose of this prospective study was to investigate the influence of insulin on nutrition, lung function and clinical status of cystic fibrosis patients. For a period of 5 y, and at 6-mo intervals, body mass index, forced expiratory volume in 1 sec, Shwachman score, intravenous glucose tolerance test and first-phase insulin response were determined in 30 cystic fibrosis patients (age range 10-35 y) with exocrine pancreatic insufficiency. During the study period, six patients (3M and 3F; age range 15-22 y) developed diabetes and required insulin therapy. The decrease of first-phase insulin response coincided with deterioration of nutritional and clinical status, which improved significantly 6 mo after the institution of insulin. CONCLUSION: Insulin, as an anabolic hormone,
could have an influence on body mass, which may affect pulmonary function and clinical condition in cystic fibrosis. It is important to identify cystic fibrosis individuals at risk of developing diabetes so that early insulin therapy is instituted.


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Neurological features in Gaucher's disease during enzyme replacement therapy.

Ono H, Fujiwara M, Ito K, Ueda H, Mizoguchi N, Sakura N.


This report describes two patients with Gaucher's disease who had unusual clinical symptoms during enzyme replacement therapy. One patient was a female with type 3 Gaucher's disease. She developed a pericardial effusion at 7 y of age, which contained many Gaucher cells despite enzyme replacement therapy. She died from neurological deterioration during enzyme replacement therapy, despite an improvement in her visceral manifestations. The other patient is a male with type 2 Gaucher's disease, who has achieved long-term survival after being supported by mechanical ventilation and enzyme replacement therapy. While on enzyme replacement therapy at the age of 4 y, he suffered a generalized cutaneous disease which was clinically diagnosed as ichthyosis. Conclusion: These cases suggest that ordinary enzyme replacement therapy is insufficient for some of the non-neurological manifestations of severe types of Gaucher's disease.


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Paediatric care. The development of children's hospices in the UK.

Overton J.


John Overton considers the contribution which hospices have made to paediatric palliative care in the UK

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Growth hormone replacement therapy in children with medulloblastoma: use and effect on tumor control.


PURPOSE: Progress has been made in the treatment of medulloblastoma, the most common childhood malignant brain tumor: However, many long-term survivors will have posttherapy growth hormone insufficiency with resultant linear growth retardation. Growth hormone replacement therapy (GHRT) may significantly improve growth, but there is often reluctance to initiate GHRT because of concerns of an increased likelihood of tumor relapse. PATIENTS AND METHODS: This study retrospectively reviewed the use of GHRT for survivors of medulloblastoma in 11 neuro-oncology centers in North America who received initial treatment for disease between 1980 and 1993 to determine its impact on disease control. A Landmark analysis was used to evaluate the relative
risk of relapse in surviving patients. RESULTS: Five hundred forty-five consecutive patients less than 15 years of age at diagnosis were identified. Six-year progression-free survival (mean +/- SD) was 40% +/- 5% in children less than 3 years of age at diagnosis compared with 59% +/- 3% for older patients. Older patients with total or near-total resections (P = .003) and localized disease at diagnosis (P < .0001) had the highest likelihood of survival. One hundred seventy patients (33% +/- 3% of the cohort) received GHRT. GHRT use varied widely among institutions, ranging from 5% to 73%. GHRT was begun a mean of 3.9 years after diagnosis, later in children younger than 3 years at diagnosis (5.4 years). By Landmark analyses, for those surviving 2, 3, and 5 years after diagnosis, there was no evidence that GHRT increased the rate of disease relapse. CONCLUSION: This large retrospective review demonstrates that GHRT is underutilized in survivors of medulloblastoma and is used relatively late in the course of the illness. GHRT is not associated with an increased likelihood of disease relapse.

Patterns of intellectual development among survivors of pediatric medulloblastoma: a longitudinal analysis.


PURPOSE: To examine two competing hypotheses relating to intellectual loss among children treated for medulloblastoma (MB): Children with MB either: (1) lose previously learned skills and information; or (2) acquire new skills and information but at a rate slower than expected compared with healthy same-age peers. PATIENTS AND METHODS: Forty-four pediatric MB patients were evaluated who were treated with postoperative radiation therapy (XRT) with or without chemotherapy. After completion of XRT, a total of 150 examinations were conducted by use of the child version of the Wechsler Intelligence SCALES: These evaluations provided a measure of intellectual functioning called the estimated full-scale intelligence quotient (FSIQ). Changes in patient performance corrected for age (scaled scores) as well as the uncorrected performance (raw scores) were analyzed. RESULTS: At the time of the most recent examination, the obtained mean estimated FSIQ of 83.57 was more than one SD below expected population norms. A significant decline in cognitive performance during the time since XRT was demonstrated, with a mean loss of 2.55 estimated FSIQ points per year (P = .0001). An analysis for the basis of the intelligence quotient (IQ) loss revealed that subtest raw score values increased significantly over time since XRT, but the rate of increase was less than normally expected, which resulted in decreased IQ scores. CONCLUSION: These results support the hypothesis that MB patients demonstrate a decline in IQ values because of an inability to acquire new skills and information at a rate comparable to their healthy same-age peers, as opposed to a loss of previously acquired information and skills.

Paediatric care. The evolution of palliative care for children in Greece.

Papadatou D.

Danai Papadatou examines how the traditional Greek family approach to dying has influenced the development of paediatric hospices.

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**Caring for dying children: A comparative study of nurses' experiences in Greece and Hong Kong.**

Papadatou D, Martinson IM, Chung PM.


The purpose of this transcultural descriptive study was to explore the subjective experiences of 63 oncology and critical care nurses who provide care to dying children in Greece and Hong Kong. Semistructured interviews were conducted with 39 Greek and 24 Chinese nurses who described their experiences and responses to the dying process and death of children. The data were analyzed qualitatively and quantitatively, and nurses' responses were compared for their work setting (oncology versus critical care) and their ethnic background (Greek versus Chinese). Findings revealed that most nurses experience a sense of helplessness when caring for a dying patient and difficulties in their communication with the child and parents during the terminal phase of the disease. The large majority acknowledge that the impending or actual death of a patient elicits a grieving process, which is characterized by a fluctuation between experiencing and avoiding loss and grief. Greek and Chinese nurses differ in their expression of their grief and how they attribute meaning to childhood death. Despite the suffering caused by multiple deaths, nurses report significant rewards from caring for chronically and acutely ill children, and the majority are satisfied with their job, despite the difficulties they encounter, in both countries, mostly as a result of shortage in personnel and cooperation problems with physicians.

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**Family stress in the intensive care unit.**

Patricia Lange M.


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**Genetic predisposition and treatment-related leukemia.**

Perentesis JP.


Treatment-related leukemias are one of the most devastating late complications of cancer therapy. Patients with rare cancer predisposition syndromes including neurofibromatosis type 1 and inherited p53 mutations are at an increased risk for this complication. Other patients may have increased susceptibility because they possess common genetic polymorphisms in drug-metabolizing enzymes that result in impaired detoxification of chemotherapy or inefficient repair of drug-induced genetic damage. We review studies that have identified a potential role for polymorphisms in the genes encoding the glutathione- S-transferases (GSTs), NAD(P) H: quinone oxidoreductase, myeloperoxidase,
N-acetyltransferase (NATs), cytochrome P450 (CYP) 1A1 and 3A4, methylenetetrahydrofolate reductase (MTHFR), cystathionine- beta-synthase (CBS), and others in the etiology of primary or secondary acute leukemias, and therapy-related complications. The identification of high risk polymorphisms and use of pharmacogenetically-guided therapies holds promise to improve the outcome of cancer therapy and reduce the risk of treatment-related leukemias.


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Palliative and terminal care for dying children; proposals for better care.

Perilongo G, Rigon L, Sainati L, Cesaro S, Carli M, Zanesco L.


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Patent ductus arteriosus and cystic periventricular leucomalacia in preterm infants.

Pladys P, Beuchee A, Wodey E, Treguier C, Lassel L, Betremieux P.


AIM: To test the association between early disturbances in hemodynamics induced by left-to-right shunting through the duct and cystic periventricular leucomalacia. PATIENTS: Forty-six preterm infants (27-32 wk) admitted to the neonatal intensive care unit with risk criteria. METHODS: Patent ductus arteriosus was evaluated on days 1 and 4, and was significant (sPDA) in cases of absent or reversed end diastolic flow in the subductal aorta. Resistance index was measured in the anterior cerebral artery and in the subductal aorta. MAIN OUTCOME: Diagnosis of cystic periventricular leucomalacia between day 10 and day 50. RESULTS: The 12 infants who developed cystic periventricular leucomalacia were compared with those who did not. On day 1, sPDA was more frequent (64% vs 26%; p = 0.03) in the cystic periventricular leucomalacia group, left ventricular output was higher (median = 341 vs 279 ml kg-1.min-1; p = 0.005), and rescue surfactant was more frequently used (83% vs 47%; p = 0.03). This latter association was confirmed by multivariate analysis. Resistance index in the anterior cerebral artery was increased in cases of significant patent ductus arteriosus (p < 0.01) and was correlated with resistance index in the subductal aorta. CONCLUSION: On day 1 in this selected population, sPDA has an effect on blood flow velocity waveform in cerebral arteries and is associated with an increase in the emergence of cystic periventricular leucomalacia. This association could be casual rather than causal.


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Toward an ethical consultation in intensive care?


No abstract available
Results of the arterial switch operation in neonates with transposed great arteries.


BACKGROUND: The arterial switch operation is judged the best palliative operation for neonates with transposed great arteries. We aimed to assess the value of analysing a large series of unselected cases by this technically demanding operation and formulate a realistic prognosis. METHODS: We reviewed all 432 neonates (mean age at operation 7 days, mean weight 3.25 kg) who underwent an arterial switch operation between 1987 and 1999. Follow-up (mean time: 4.9 years) was complete in 412 patients. FINDINGS: Survival probability and freedom from reoperation was 94% and 78% at 10 years, respectively. 26 patients died, 16 because of myocardial ischaemia. Risk factors for death included early experience, low weight, associated cardiovascular malformations (especially hypoplasia of the right ventricle or aortic arch), and difficult patterns of coronary arteries. The risk of the coronary artery pattern was greatly reduced in those who had recent operations. At last follow-up, 90% of patients had normal life without treatment, and 94% a normal heart function on echocardiography. INTERPRETATION: The arterial switch operation in neonates achieves excellent results mid-term. Obstruction of the translocated coronary arteries is responsible for most deaths and a substantial number of reoperations. Although confirmation is needed, these results allow anticipation of a favourable long-term prognosis.


Caring for Bereaved Patients: "All the Doctors Just Suddenly Go". [Miscellaneous Article].

Prigerson HG, Jacobs SCM.


Despite the frequency with which physicians encounter bereaved patients, medical training offers little guidance in the provision of bereavement ("after") care. Physicians are often uncertain of how to distinguish between normal and pathological grief reactions in their bereaved patients, and how to manage their health care. Bereavement is associated with declines in health, inappropriate health service use, and increased risk of death. Identifying and intervening on behalf of bereaved patients could help address those increased risks. We examine the experience of a woman widowed for 2 years to illustrate distinctions between symptoms and outcomes of uncomplicated and complicated grief, recommend approaches to physician interactions with bereaved patients, and offer guidelines for professional intervention in aftercare., *JAMA*.2001;286:1369-1376, Copyright 2001 by the American Medical Association. All Rights Reserved. Applicable FARS/DFARS Restrictions Apply to Government Use. American Medical Association, 515 N. State St, Chicago, IL 60610.

Nutritional support at home and in the community.

Puntis JW.

Technical developments in feeding, together with the growth of support structures in the community has lead to a steady increase in the number of children receiving home enteral tube feeding and home parenteral nutrition. In many cases the adverse nutritional consequences of disease can be ameliorated or prevented, and long term parenteral nutrition represents a life saving intervention. Careful follow up of children receiving home nutritional therapy is necessary to establish the ratio of risks to benefits. A considerable burden is sometimes placed on family or other carers who therefore require adequate training and ongoing support. The respective responsibilities of different agencies relating to funding and support tasks require more clear definition.

http://www.archdischild.com/cgi/content/full/archdischild;84/4/295
http://www.archdischild.com/cgi/content/abstract/archdischild;84/4/295

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Patients' needs at the end of life.

Rabow MW, McPhee SJ.


http://www.jco.org/cgi/content/full/19/15/3585

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How and when deciding to stop curative treatments in pediatric oncology?

Raimondo G, Hartmann O.


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Torticollis revealing as a symptom of acute lymphoblastic leukaemia in a fourteen-month-old girl.

Rauch R, Jungert J, Rupprecht T, Greil J.


The differential diagnosis of acquired torticollis in infants and children comprises a large variety of disorders, including inflammation, neurologic disease or trauma. CONCLUSION: Spinal cord tumours in patients with ALL are a rarely reported entity and secondary spinal involvement in leukaemia presenting as head tilt is even more uncommon.


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Analysis of handwriting of children during treatment for acute lymphoblastic leukemia.

Reinders-Messelink HA, Schoemaker MM, Snijders TA, Goeken LN, Bokkerink JP, Kamps WA.


BACKGROUND: Children treated for acute lymphoblastic leukemia (ALL) often complain about handwriting problems. PROCEDURE: Using a computerized writing task, we have prospectively studied the processes necessary for the production of handwriting movements in 11 children (5-12 years old) during treatment for ALL. Children were tested at time points closely related to the vincristine administration. RESULTS AND CONCLUSIONS: Children treated for ALL drew slower, with longer pause durations and increased drawing pressure. Children were able to overcome the problems, except for a consistently increased drawing pressure. This increased drawing pressure may be an attempt of the children to obtain sufficient kinesthetic information and thus can be seen as an adequate adaptation mechanism in case of peripheral neuropathy due to the neurotoxic effects of vincristine. However, neurotoxic effects of other cytostatic drugs cannot be excluded.


Precocious puberty in children with tumours of the suprasellar and pineal areas: organic central precocious puberty.

Rivarola, Belgorosky A, Mendilaharzu H, Vidal G.


During the past 11 y, 115 children younger than 8/9 y of age (female/male) with tumours of the suprasellar or pineal areas were followed in our clinic to study the incidence of precocious puberty. In addition, type of central lesion, clinical characteristics and gonadotropic secretion were studied in order to elucidate the different mechanisms of gonadal activation. A control group of 21 patients with idiopathic precocious puberty and a control group of 10 age-matched patients with suprasellar tumours without precocious puberty were also studied. Precocious puberty associated with organic central lesions was found at diagnosis in 30 patients (26%), in 9 out of 48 patients with glial cell tumours (18.7%), 6 out of 9 patients with germ cell tumours (66.6%), 11 out of 11 patients with hypothalamic hamartomas (100%) and in 4 out of 4 patients with subarachnoid cysts or arachnoidocele (100%). Precocious puberty was not found in any of 36 patients with craniopharyngioma. With the exception of one patient with pineal germinoma, all lesions were localized to the suprasellar area. In all patients with hypothalamic hamartoma, precocious puberty was diagnosed before 4 y of age, while in most patients with the other lesions, it was diagnosed after this age. Height SDS, weight increase and advancement of bone age were similar in both idiopathic and organic central precocious puberty. Maximal LH responses to GnRH in idiopathic and organic central precocious puberty were similar except for germ cell tumours. Patients with suprasellar tumours without precocious puberty had lower maximal LH (but not FSH) responses to GnRH, with the exception of germ cell tumours. In the latter, elevation of serum beta-hCG indicates that this gonadotropin was responsible for gonadal stimulation. In hypothalamic hamartomas, the prepubertal hiatus in the activity of the GnRH pulse generator was absent. The mechanism of this failure in the inactivation of GnRH is unknown. Data suggest that in glial cell tumours and in subarachnoid cysts, an unknown factor, probably secreted by the tumours, advances the tempo of GnRH maturation. Therefore, the aetiology of organic central precocious puberty is multiple and is directly related to location and type of lesion. CONCLUSION: This clinical information suggests that the onset of puberty is not the result of the disruption of a putative pulse generator inhibitory influence but the consequence of secretion of stimulatory substances by the lesions.

Acknowledging the extra care parents give their disabled children.

Roberts K, Lawton D.


Around 150,000 families in the UK care for a severely disabled child under the age of 16. Many of these families receive assistance from the Family Fund Trust, which provides grants and information relating to the care of a severely disabled child. The aim of this study was to identify patterns of extra care needs among severely disabled children known to the Trust. Extra care needs are requirements for care not experienced by similarly aged non-disabled children. The research comprised analysis of 40,000 records from the Trust database and qualitative exploration of the extra care needs of disabled children with parents and Trust staff. Although all children require parenting, the care parents give disabled children generally exceeds that given to a non-disabled child. Quantitative analysis showed that the majority of children in the sample required extra assistance or supervision with multiple areas of daily life. With each of five activities (washing, dressing, meal times, during the night and keeping occupied), >70% of children needed extra help and, on average, each child needed extra help or supervision in six areas of daily life. Cluster analysis indicated distinctive combinations of extra care needs. Qualitative material indicated variety in extra care tasks undertaken (physical help, supervision, guidance) and causal factors (physical limitations, cognitive difficulties, behavioural problems). The findings confirm that severely disabled children have considerable extra care needs in many areas of daily life. Parents want professionals to recognize and offer explicit acknowledgement of the extra care they give their disabled children.


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Cystic fibrosis-related diabetes mellitus: clinical impact of prediabetes and effects of insulin therapy.


In patients with cystic fibrosis (CF), glucose intolerance preceding diabetes (prediabetes) may have adverse effects on nutritional status and respiratory function, which are reversible after the start of insulin therapy. Respiratory function (forced vital capacity and forced expiratory volume in one second) and body mass index (BMI) were compared retrospectively in a French cohort of 14 patients during the 5 y preceding insulin therapy for diabetes and in 14 age- and sex-matched controls with normal oral glucose tolerance tests. In the diabetic group, all three parameters deviated increasingly from the values in the controls; the differences became statistically different during the 6 mo before insulin therapy. The effect was more important in patients for whom diabetes mellitus was diagnosed on the basis of symptoms of hyperglycaemia than in patients for whom it was diagnosed by systematic screening, but still present in the latter. After insulin was started, respiratory function improved and the BMI returned to normal within 1 y. The annual insulin requirement increased from 0.62 during the first year to 1.25 during the fifth year. Glycosylated haemoglobin (HbA1c) values ranged from 6.6 to 7.8%. Only 2 episodes of severe hypoglycaemia were recorded over 42 patient-years of follow-up. The insulin regimen most often used was two daily injections of a mixture of short- and intermediate-acting insulin (n = 10) given with an insulin pen.

CONCLUSION: The clinical status of CF patients who will need insulin therapy deteriorates before the start of insulin. In patients with CF-related diabetes, with or without fasting hyperglycaemia, insulin therapy improves anabolism and provides good glycaemic control with few severe hypoglycaemic episodes.

Defining a pulmonary exacerbation in cystic fibrosis.


**OBJECTIVES:** Despite the central importance of pulmonary exacerbations (PExs) as an outcome measure in cystic fibrosis clinical trials, no standardized definition of PEx exists. We conducted a prospective, multicenter study to establish a standardized PEx definition and score for use in clinical trials, based on clinical status rather than on treatment decisions. **STUDY DESIGN:** Subjects were 246 patients enrolled in the placebo arm of a randomized, controlled trial of tobramycin for inhalation. Physician-investigators completed PEx questionnaires on all subjects at scheduled intervals during the 6-month study, indicating new or worsening symptoms, physical examination findings, and impression of PEx status (presence or absence and severity). Logistic regression was used to assess the relative importance of each of the characteristics in predicting a PEx. **RESULTS:** We developed 2 PEx scores that use easily ascertained symptoms and chest examination findings; one also includes change in forced expiratory volume in 1 second over the preceding month. Both scores were sensitive and specific for predicting the presence of a PEx (sensitivity, 86%; specificity, 86%). The scores were validated in subjects in the intervention arm of the trial. **CONCLUSION:** We hope that the proposed PEx score might serve as a standardized outcome measure for future clinical trials in cystic fibrosis, allowing meaningful comparisons of study results.

http://www.mosby.com/scripts/om.dll/serve?action=searchDB&searchDBfor=art&artType=abs&id=a117288&target=

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Childhood cancer etiology: recent reports.

Ross JA, Davies SM.


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Caring for the dying: why is it so hard for physicians?

Rousseau P.


http://www.ewjm.com/cgi/content/full/175/4/284

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Kindness and the end of life.
End-of-life care in the intensive care unit: a research agenda.

Rubenfeld GD, Randall Curtis J.


BACKGROUND: The intensive care unit (ICU) represents a unique clinical setting in which mortality is relatively high and the professional culture tends to be one of "rescue therapy" using technological and invasive interventions. For these reasons, the ICU is an important environment for understanding and improving end-of-life care. Although there have been consensus statements and review articles on end-of-life care in the ICU, there is limited evidence on which to base an assessment of best practices for providing high-quality end-of-life care in this setting.

OBJECTIVE: To convene a Working Group of experts in critical care, palliative medicine, medical ethics, and medical law to address the question "What research needs to be done to improve end-of-life care to patients in the ICU?"

METHODS: Participants were identified for membership in the Working Group by purposive sampling within the fields of critical care medicine and nursing, palliative medicine, and medical ethics; others were chosen to represent social work and hospital chaplains. Through a process of breakout and plenary sessions, the group identified important questions that need to be addressed in the areas of defining the problem, identifying solutions, evaluating solutions, and overcoming barriers.

CONCLUSIONS: Outlining unanswered questions on end-of-life care in the ICU is a first step to providing the answers that will allow us to improve care to patients dying in the ICU. These questions also serve to focus clinicians and educators on the important areas for improving quality of care.

Opsoclonus-myoclonus-ataxia syndrome in neuroblastoma: clinical outcome and antineuronal antibodies-a report from the Children's Cancer Group Study.


BACKGROUND: Opsoclonus-myoclonus-ataxia (OMA) is a paraneoplastic neurologic syndrome affecting 2-3% of children with neuroblastoma. Although children with OMA and neuroblastoma may have higher survival, many experience a significant amount of late neurologic impairment, which may be immunologically mediated. The aim of this study was to compare the outcome of neuroblastoma patients with and without OMA, relating to prognostic factors, treatment, and the presence or absence of anti-neuronal antibodies. PROCEDURE: Questionnaires were mailed out requesting information on the current neurologic status of patients who submitted sera at diagnosis to the Children’s Cancer Group serum bank from 1980 to 1994. Information was requested on clinical and biological patient characteristics as well as clinical aspects of the patients identified as having OMA syndrome, including presentation and treatment for OMA, late sequelae of OMA, the presence or absence of antineuronal antibodies, and survival. Sera from 16 of the OMA patients and 48 case-controls with neuroblastoma were assayed for anti-
neuronal antibodies. RESULTS: Of the 675 responses received, 21 patients had OMA. Ninety percent of OMA patients presented with non-metastatic disease, vs. 35% of non-OMA patients. Estimated 3-year survival for the OMA patients with nonmetastatic disease (stage I, II, III) greater than 1 year of age was 100% vs. 77% for similar non-OMA patients (P = 0.0222). At follow-up, 14/19 evaluable OMA patients displayed some form of developmental or neurologic abnormality. There was no significant correlation of late sequelae with antineuronal antibodies, age, time between OMA symptoms and diagnosis, or treatment given for tumor or OMA. There was a significant correlation of late sequelae with lower stage disease (I and II) compared to more advanced disease (III and IV). CONCLUSIONS: Patients with OMA and neuroblastoma have excellent survival but a high risk of neurologic sequelae. Favorable disease stage correlates with a higher risk for development of neurologic sequelae. The role of anti-neuronal antibodies in late sequelae of OMA needs further clarification.


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Microcephalus, medulloblastoma and excessive toxicity from chemotherapy: an unusual presentation of Fanconi anaemia.

Ruud E, Wesenberg F.


Fanconi anaemia is a genetically and phenotypically heterogeneous disorder with different forms of clinical presentation. In this case the patient had suffered from microcephalus and delayed motor development from birth, but extensive investigation did not disclose any aetiology. At 3.5 y she developed a cerebellar medulloblastoma which was treated with surgery and chemotherapy. Following chemotherapy with alkylating agents she suffered from severe bone marrow aplasia which caused life-threatening infections, feeding problems and impaired kidney function. Fanconi anaemia was suspected, but it took 2 mo before the chromosome fragility test came out positive. From the moment diagnosis of Fanconi anaemia was made, no further active treatment was given. The patient's condition improved for some time, but she relapsed and died exactly 1 y after the first diagnosis of brain tumour. CONCLUSION: Fanconi anaemia must always be suspected in patients who experience excessive toxicity from chemotherapy regardless of the type of malignancy and congenital malformations.


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No disadvantage in outcome of using matched unrelated donors as compared with matched sibling donors for bone marrow transplantation in children with acute lymphoblastic leukemia in second remission.


PURPOSE: We evaluated the outcome of children with acute lymphoblastic leukemia (ALL) in second remission (2CR), comparing bone marrow transplantation (BMT) using either matched sibling donors or unrelated donors (URDs). PATIENTS AND METHODS: A total of 65 patients, aged 2 months to 20 years at BMT, with ALL in 2CR underwent allogeneic BMT at seven Nordic centers during 1990 to 1997. Of the first relapses, 85% were in bone marrow; 46% occurred on therapy, and 54%, off therapy. The preparative regimens were cyclophosphamide plus total-body irradiation +/- antithymocyte/antilymphocyte globulin, busulfan plus cyclophosphamide +/-...
antithymocyte/antilymphocyte globulin, or cytarabine plus total-body irradiation. Of the allografts, 37 were from HLA-matched siblings and 28 were from URDs. RESULTS: In the sibling versus URD graft recipient groups, the posttransplantation 5-year event-free survival was 39% versus 54% (P = .4), the estimated posttransplantation relapse rate was 76% versus 40% (P = not significant [NS]), and the toxic death rate was 19% versus 11% (P = NS). The incidence of significant (grade 2 to 4) acute graft-versus-host disease (GVHD) was 38% versus 64% (P < .05) and was 14% versus 32% (P < .10) for severe (grade 3 to 4) acute GVHD; the incidence of chronic GVHD was 26% versus 57% (P < .05) and was 13% versus 22% (P = NS) for extensive chronic GVHD in the sibling and URD groups. CONCLUSION: BMT with matched URD allografts offers at least equal survival for children with ALL in 2CR, as compared with allografts from matched sibling donors. URD allografts were not associated with a higher toxic mortality rate, although both acute and chronic GVHD were more frequent with URD. Indications for using matched URD allografts in ALL 2CR can be considered the same as for using matched sibling donors.

http://www.jco.org/cgi/content/full/19/14/3406
http://www.jco.org/cgi/content/abstract/19/14/3406

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Complications of cervical spine surgery.

Salcman M.


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Prospective, multicenter evaluation of risk factors associated with invasive bacterial infection in children with cancer, neutropenia, and fever.


PURPOSE: To identify clinical and laboratory parameters present at the time of a first evaluation that could help predict which children with cancer, fever, and neutropenia were at high risk or low risk for an invasive bacterial infection. PATIENTS AND METHODS: Over a 17-month period, all children with cancer, fever, and neutropenia admitted to five hospitals in Santiago, Chile, were enrolled onto a prospective protocol. Associations between admission parameters and risk for invasive bacterial infection were assessed by univariate and logistic regression analyses. RESULTS: A total of 447 febrile neutropenic episodes occurred in 257 children. Five parameters were statistically independent risk factors for an invasive bacterial infection. Ranked by order of significance, they were as follows: C-reactive protein levels of 90 mg/L or higher (relative risk [RR], 4.2; 95% confidence interval [CI], 3.6 to 4.8); presence of hypotension (RR, 2.7; 95% CI, 2.3 to 3.2); relapse of leukemia as cancer type (RR, 1.8, 95% CI, 1.7 to 2.3); platelet count less than or equal to 50,000/mm(3) (RR, 1.7; 95% CI, 1.4 to 2.2); and recent (< or = 7 days) chemotherapy (RR, 1.3; 95% CI, 1.1 to 1.6). Other previously postulated risk factors (magnitude of fever, monocyte count) were not independent risk factors in this study population. CONCLUSION: In a large population of children, common clinical and laboratory admission parameters were identified that can help predict the risk for an invasive bacterial infection. These results encourage the possibility of a more selective management strategy for these children.

http://www.jco.org/cgi/content/full/19/14/3415
Chronic lung disease: oxygen dogma revisited.

Saugstad OD.


Since the discovery of retrolental fibroplasia, and the role of oxygen in its development, oxygen has been considered a double-edged sword in neonatal medicine, the utmost care being exercised in order not to give too much oxygen (1). However, the important observation that hypoxaemia might induce pulmonary vasoconstriction (2) and airway constriction (3) in infants at risk for bronchopulmonary dysplasia has resulted in only a minor upward adjustment of oxygen supplementation in many neonatal units. Since oxygen toxicity has long been linked not only to retinopathy of prematurity but also to bronchopulmonary dysplasia (4), it is relevant to ask whether an increased FiO2 might have any detrimental effects on babies.


Scheinberg A, O'Flaherty S, Chaseling R, Dexter M.


OBJECTIVES: To determine whether continuous intrathecal baclofen infusion (CIBI) would decrease spasticity and improve function in children with spastic cerebral palsy. DESIGN: Prospective study with measurement of changes from baseline assessed at regular intervals for 6 months following the intervention. PATIENTS: Two subjects aged 8 and 9 years with cerebral palsy. Inclusion criteria included severe spasticity and age greater than 4 years. STUDY CENTRE: Department of Rehabilitation, The Children's Hospital at Westmead, Westmead, New South Wales, Australia. RESULTS: Both subjects had clinically significant reductions in lower limb spasticity from a single intrathecal dose of baclofen (screening procedure), and had intrathecal pumps implanted. An intensive physical therapy programme was provided for both subjects in the 1-3-month period after commencing CIBI, in order to maximize functional gains. Modified Ashworth Scale scores remained reduced during the 6-month study period. There was a clinically significant increase in upper limb function for Subject 1. Gross Motor Function Measure scores decreased from 22% to 19% for Subject 1, and increased from 6% to 10% for Subject 2 over the study period. Paediatric Evaluation of Disability Inventory scores for Subject 1 showed a reduction in the level of caregiver assistance required, while Subject 2 showed significant improvement in the functional mobility domain. The major changes noted in the parent questionnaires were reduction in tone, increased range of motion and reduced time taken helping with activities of daily living such as toilet and dressing. No significant side-effect was seen after the screening procedure or after continuous intrathecal infusion. Both subjects' parents felt their child was improved following the intervention. CONCLUSIONS: Children with spastic cerebral palsy can have their spasticity effectively reduced with CIBI. In this study of two children, the clinical improvements were encouraging, and it is proposed that CIBI may be of benefit to those patients whose level of spasticity severely interferes with function. Further studies using multidimensional assessment approaches, with larger numbers of children, are warranted.
Online personal medical records: are they reliable for acute/critical care?

Schneider JH.


OBJECTIVE: To provide an introduction to Internet-based Online Personal Medical Records (OPMRs), to assess their use and limitations in acute/critical care situations, and to identify potential improvements that could increase their usefulness. DESIGN: A review of publicly available Internet-based OPMRs conducted in April 2001. DATA SOURCES: Twenty-nine OPMR sites were identified in March 2000 using ten Internet search engines with the search term "Personal Medical Records." Through 2000 and 2001, an additional 37 sites were identified using lists obtained from trade journals and through the author's participation in standards-setting meetings. MEASUREMENTS: Each publicly available site was reviewed to assess suitability for acute/critical care situations using four measures developed by the author and for general use using eight measures developed in a standards-setting process described in the article. RESULTS: Of the 66 companies identified, only 16 still offer OPMRs that are available to the public on the Internet. None of these met all of the evaluation measures. Only 19% had rapid emergency access capabilities and only 63% provided medical summaries of the record. Security and confidentiality issues were well addressed in 94% of sites. Data portability was virtually nonexistent because all OPMRs lacked the ability to exchange data electronically with other OPMRs, and only two OPMRs permitted data transfer from physician electronic medical records. Controls over data accuracy were poor: 81% of sites allowed entry of dates for medical treatment before the patient's date of birth, and one site actually gave incorrect medical advice. OPMRs were periodically inaccessible because of programming deficiencies. Finally, approximately 40 sites ceased providing OPMRs in the past year, with the probable loss of patient information. CONCLUSIONS: Most OPMRs are not ready for use in acute/critical care situations. Many are just electronic versions of the paper-based health record notebooks that patients have used for years. They have, however, great promise and, with further development, could form the basis of a new medical record system that could contribute to improving the quality of medical care.


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Guiding your child through grief.

Schonfeld DJ.


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Ethical Decisions Regarding Nutrition and the Terminally Ill. [Article].

Schwarte AMS.

*Gastroenterology Nursing 2001;24(1):29-33.*

Care of the terminally ill is rewarding and challenging. Nurses have the potential to affect the quality of these patients' lives. A recurrent theme voiced by terminally ill patients and their families is the nutritional ramifications associated with the cessation of nutritional intake. Various ethical perspectives provide a useful guide for decision making when determining whether fluids and nutrition should be continued in the terminally ill. This article will discuss various ethical perspectives in relation to nutrition cessation in the terminally ill. Basic physiologic changes
that occur in the terminal patient will also be addressed. Nurses play an important role in insuring quality of care, particularly in supporting patients and families as they make decisions regarding nutritional management at the end of life., (C) The Society of Gastroenterology Nurses & Associates 2001. All Rights Reserved.

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Optimizing treatment for chronic congestive heart failure in children.

Shaddy RE.


Treatment strategies for chronic heart failure in children have generally been extrapolated from studies in adults with heart failure. This presentation reviews the existing knowledge and recommendations regarding the treatment of chronic heart failure in adults and the information that is available in children. Medications currently recommended for use in adults include diuretics, digoxin, angiotensin-converting enzyme inhibitors, and beta-blockers. These recommendations are based on results from large, randomized, multicenter trials. Anecdotal evidence suggests similar beneficial effects of these medications in children. The fact that the etiologies, pathophysiology, and physiologic consequences of heart failure in children often differ greatly from those in adults, however, justifies the development of prospective, randomized trials to evaluate these medications specifically in children. Findings from these types of studies will provide critical information for developing guidelines for the appropriate treatment of children with chronic heart failure.


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Urinary calcium excretion in enterally fed disabled children.

Shetty AK, Carlin J, Mughal MZ.

_Arch Dis Child 2001;85(1):58-9._

The urine calcium/creatinine (Ca/Cr) ratio was measured in 17 enterally tube fed disabled children. Urine Ca/Cr ratios were inversely related to dietary calcium intake in immobile children (r = -0.57, p < 0.05) but not in those who were able to stand or walk (mobile children) (r = 0.4, p = 0.51). None of the subjects had evidence of nephrocalcinosis or renal calculi detectable by renal ultrasonography.

http://www.archdischild.com/cgi/content/full/archdischild;85/1/58
http://www.archdischild.com/cgi/content/abstract/archdischild;85/1/58

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Late mortality in childhood cancer: two excellent studies bring good news tempered by room for improvement.

Simone JV.


A simple stratification factor prognostic for survival in advanced cancer: the good/bad/uncertain index.


PURPOSE: This article summarizes the third step of a research program to identify variables that supplement the predictive power of the Eastern Cooperative Oncology Group (ECOG) performance status (PS) for survival. The objective was to produce a simple, practical, stratification factor for phase III oncology clinical trials involving patients with advanced malignant disease. PATIENTS AND METHODS: A questionnaire was administered to 729 patients with metastatic colorectal or lung cancers. Patients provided a Karnofsky index and appetite rating while physicians provided a survival estimate and the ECOG-PS. Scores for each item were categorized as having a positive, neutral, or negative indication for survival. A patient was classified as having a relatively good prognosis if three or more of the four items showed a positive indication, a bad prognosis if three or more items were negative, and an uncertain prognosis otherwise (Good/Bad/Uncertain [GBU] index). RESULTS: The GBU index improved on the prognostic power of a Cox model quartile index and PS alone and increased the accuracy of survival classification estimates by 5% to 10% more than ECOG-PS alone. For patients with PS of 0 or 1, significant survival patterns exist between GBU groups (P=.002 and .0001, respectively). CONCLUSION: The GBU index may be recommended as a supplementary stratification factor for certain future phase III trials in metastatic lung or colorectal cancer where patient heterogeneity is a particular concern. The GBU represents a relatively modest increase to the cost and patient burden of a clinical trial given the additional control that is achieved over the potentially confounding concomitant to the treatment variable.

Hepatic storage of glycogen in Niemann-Pick disease type B.

Smith WE, Kahler SG, Frush DP, Milov DE, Gottfried MR, Chen YT.


We report 2 patients with confirmed Niemann-Pick disease, type B, with previous diagnoses of glycogen storage disease based on excessive glycogen on liver biopsy specimens. These cases emphasize the importance of a complete evaluation, including biochemical confirmation, for patients with suspected metabolic storage diseases.

Bony morbidity in children treated for acute lymphoblastic leukemia.
PURPOSE: Corticosteroids are widely used in the treatment of acute lymphoblastic leukemia (ALL). To determine the frequency of corticosteroid-associated bony morbidity in children with ALL, we retrospectively evaluated the incidence of fractures and osteonecrosis (ON) on two consecutive pediatric ALL protocols. PATIENTS AND METHODS: One hundred seventy-six consecutive children were treated for ALL between 1987 and 1995 at the Dana-Farber Cancer Institute and Children's Hospital. Prednisone was used as the corticosteroid during postremission therapy from 1987 to 1991, and dexamethasone was used from 1991 to 1995. Medical records for all patients were reviewed to assess the occurrence of fractures and ON. RESULTS: With a median follow-up of 7.6 years, the 5-year cumulative incidence (CI) +/- SE of any bony morbidity for the 176 patients was 30% +/- 4%, with a 5-year CI of fractures of 28% +/- 3% and of ON of 7% +/- 2%. With multivariate analysis, independent predictors of bony morbidity included age 9 to 18 years at diagnosis (P <.01), male sex (P <.01), and treatment with dexamethasone (P =.01). Dexamethasone was associated with a higher risk of fractures (5-year CI, 36% +/- 5% v 20% +/- 4% with prednisone; P =.04), but not ON (P =.40). The 5-year event-free survival for the 176 patients was 79% +/- 3%. CONCLUSION: Children treated for ALL had a high incidence of fractures and ON. Older children, boys, and patients receiving dexamethasone were at increased risk for the development of bony morbidity. Future studies should attempt to minimize corticosteroid-associated bony morbidity without compromising clinical efficacy.

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When children have to die.


Archives De Pedriatrie 2001;8(7):751-753.

Palliative care has recently been developed in adults. However, children die as well and quite a few articles have been published on this special issue. The object of this article is to summarize the available literature in order to call for the development of clinical policies and minimum standards adapted to French pediatrics. (C) 2001 Editions scientifiques et medicales Elsevier SAS.

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Heart failure in pediatric septic shock: utilizing inotropic support.

Tabbutt S.  


Septic shock presents a unique challenge in the pediatric patient. Sepsis stimulates the release of inflammatory mediators that can compromise cardiac function. Oxygen extraction abnormalities, diminished responses to adrenergic agonists, and impaired ventricular function often result. After fluid resuscitation and antibiotic therapy, careful cardiovascular assessment is needed to administer appropriate inotropic and vasoactive drugs.
Lung function measurements in young children with spinal muscle atrophy; a cross sectional survey on the effect of position and bracing.

Tangsrud SE, Carlsen KC, Lund-Petersen I, Carlsen KH. 


BACKGROUND: Spinal muscular atrophy (SMA) affects respiratory muscles, which in addition to progressive scoliosis leads to respiratory impairment. Children with developing scoliosis are usually treated with spinal bracing to delay the progress. AIMS: To assess the impact of body position and application of spinal bracing on lung function during tidal breathing in children with SMA. METHODS: Lung function was determined by tidal flow volume loops and passive respiratory mechanics (single breath occlusion technique) in all eight children in southern Norway with SMA type I and II, in both the sitting and supine position. Additional measurements were performed with and without bracing in five children. Muscle strength was assessed by the Brooks scale. RESULTS: Tidal expiratory volume (V(E)) and compliance of the respiratory system (CRS) tended to be higher in the sitting compared to the supine position, but this was not statistically significant. However, applying bracing in the sitting position significantly reduced V(E). The highest values of CRS and V(E) were found in the sitting position without bracing. CONCLUSION: Impairment of tidal respiration must be considered when applying spinal bracing in very young children developing scoliosis with SMA.

Enteral nutrition: how do we get more of the good and less of the bad and ugly?

Thorborg P.


Optimal duration of preoperative therapy in unilateral and nonmetastatic Wilms' tumor in children older than 6 months: results of the Ninth International Society of Pediatric Oncology Wilms' Tumor Trial and Study.


PURPOSE: To determine the optimal duration of preoperative chemotherapy to further increase the proportion of stage I tumors by comparison of two regimens in the treatment of patients older than 6 months who have unilateral Wilms' tumor. PATIENTS AND METHODS: Eligible patients (n = 382) initially received four weekly doses of
vincristine (VCR) and two courses of actinomycin D (AMD) and were randomized either to be operated on (4-week group \([n = 193]\)) or to receive 4 more weeks of the same chemotherapy regimen (8-week group \([n = 189]\)). The assessment criterion was the observed percentage of stage I tumors. After surgery, patients were assigned according to tumor stage and histology to four different treatment groups: stage I and favorable histology \((n = 5)\) were to have no further treatment (NFT); stage I and standard histology or anaplasia \((n = 244)\), VCR and AMD for 17 weeks (AV); stages II and III and favorable or standard histology, VCR, AMD, and an anthracycline for 27 weeks (AVE) with no abdominal radiotherapy for stage II N0 disease \((n = 75)\) or with a 15-Gy dose of abdominal irradiation (RTH) in case of stages IIN1 and III \((n = 56)\). Anaplastic tumors staged higher than I or clear-cell sarcoma of the kidney \((14)\), AMD, VCR, an anthracycline, and ifosfamide for 36 weeks (DEVI). RESULTS: No advantage was found in favor of prolonged preoperative treatment. The percentages obtained for the 4-week and the 8-week groups, respectively, were as follows: stage I, 64\% versus 62\%; intraoperative tumor rupture rate, 1\% versus 3\%; 2-year EFS, 84\% versus 83\%; and 5-year OS, 92\% versus 87\%. Two-year EFS and 5-year OS rates, respectively, of the different treatment groups were as follows: NFT, 100\% for both EFS and OS; AV, 88\%and 93\%; AVE, 84\% and 88\%; AVERTH, 71\% and 85\%; and DEVI, 71\% and 71\%. The rate of abdominal recurrences in stage II N0 nonirradiated patients was 6.6\%. CONCLUSION: The 4-week schedule pre-nephrectomy chemotherapy regimen should be considered the standard treatment. Clinical trials should continue to improve the cure rate of high-risk patients and the quality of life of children with a more favorable prognosis.

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Repeated achilles tendinitis after high dose methotrexate.

Toverud EL, Landaas S, Hellebostad M.


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Prediction of health behaviors in pediatric cancer survivors.

Tyc VL, Hadley W, Crockett G.


BACKGROUND: It is important to understand the factors that motivate survivors of childhood cancer to engage in healthy behaviors. This is because of their susceptibility to adverse late effects from their malignancy and its treatment. We specifically examined sociodemographic and health perception variables as predictors of health-protective behaviors in pediatric cancer survivors. PROCEDURE: Forty-six patients, ages 10–18 years, who were previously treated for cancer and were 1–4 years off-therapy, were assessed using a battery of testing instruments. RESULTS: Preadolescent and adolescent cancer survivors reported moderately frequent practice of a variety of health-protective behaviors. The prevalence of risky health behaviors, as indicated by alcohol and tobacco use, was low. Regression analyses indicated that the practice of health-protective behaviors was best predicted by patient's age and socioeconomic status (SES); younger adolescents and patients from higher SES more frequently engaged in healthy behaviors. Health perception variables were not significantly related to health-protective behaviors. This was true despite findings that survivors perceived themselves to be vulnerable to health problems, identified a need to protect their health, and perceived their health outcomes to be largely determined by their own behaviors. CONCLUSIONS: Sociodemographic factors should be kept in mind when designing interventions to promote
continuing good health for young cancer survivors. Also to be included is the assessment of specific health risks secondary to the cancer therapies given. Risk counseling that recognizes and builds on these variables will be most effective in helping this patient population observe sound health habits.


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Natural history of cardiovascular manifestations in Marfan syndrome.

van Karnebeek CD, Naeff MS, Mulder BJ, Hennekam RC, Offringa M.

Arch Dis Child 2001;84(2):129-37.

AIMS: To investigate the natural history of mitral valve and aortic abnormalities in patients with Marfan syndrome during childhood and adolescence. METHODS: Fifty two patients with Marfan syndrome were followed for a mean of 7.9 years. Occurrence of adverse cardiovascular outcomes was measured clinically and by ultrasound examination. RESULTS: Mitral valve prolapse (MVP) was diagnosed in 46 patients at a mean age of 9.7 years, more than 80% of whom presented as "silent MVP". Mitral regurgitation (MR) occurred in 25 patients, aortic dilatation in 43, and aortic regurgitation (AR) in 13. Both MVP and aortic dilatation developed at a constant rate during the age period 5-20 years. In 23 patients MVP was diagnosed before aortic dilatation, in 18 the reverse occurred, and in 11 patients the two abnormalities were diagnosed simultaneously. During follow up, 21 patients showed progression of mitral valve dysfunction; progression of aortic abnormalities occurred in 13. Aortic surgery was performed in 10; two died of subsequent complications. Mitral valve surgery was performed in six. In sporadic female Marfan patients the age at initial diagnosis of MVP, MR, aortic dilatation, and AR was lowest, the grade of MR and AR most severe, the time lapse between the occurrence of MVP and subsequent MR as well as between dilatation and subsequent AR shortest, and the risk for cardiovascular associated morbidity and mortality highest. CONCLUSIONS: During childhood and adolescence in Marfan syndrome, mitral valve dysfunction as well as aortic abnormalities develop and progress gradually, often without symptoms, but may cause considerable morbidity and mortality by the end of the second decade, especially in female sporadic patients.

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Tube feeding: new life for an old procedure.

Van Way CW, 3rd.


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Nocturnal oximetry in infants with cystic fibrosis.

Villa MP, Pagani J, Lucidi V, Palamides S, Ronchetti R.

Arch Dis Child 2001;84(1):50-54.
AIM: To investigate whether children with cystic fibrosis under 3 years of age have disordered breathing and episodes of oxygen desaturation during sleep. METHODS: We studied 19 infants (9 boys and 10 girls) with cystic fibrosis, mean age 13.1 months (range 3-36 months) and 20 age and sex matched healthy subjects. Patients and controls underwent an overnight polysomnographic study and respiratory function testing on the following morning. RESULTS: Seven patients with ongoing respiratory tract inflammation had disordered breathing and episodes of oxygen desaturation during sleep. Pulse oximetry showed a significantly lower mean oxygen saturation (SaO(2)) and a higher percentage of total sleep time spent with SaO(2) less than 93% in symptomatic children than in controls. CONCLUSION: Results suggest that infants and young children with cystic fibrosis and mild airways inflammation (rhinitis, cough, red throat) have episodes of oxygen desaturation during sleep.

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Discussing do-not-resuscitate status.

von Gunten CF.


http://www.jco.org/cgi/content/full/19/5/1576

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Does continuity of care matter? No: discontinuity can improve patient care.

Wachter RM.


http://www.ewjm.com/cgi/content/full/175/1/5

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Children in need... palliative care for children.

Wates S.


Paediatric palliative care is a demanding but rewarding role. Sue Wates reports.

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PaedPalLit Vol.1 No. 1 (Jan 2000 – Dec 2001)

The Internet, the electronic medical record, the pediatric intensive care unit, and everything.

Weigle CG, Markovitz BP, Pon S.

*Crit Care Med* 2001;29(8 Suppl): N166-76.

This article details how computers have changed life for those of us in pediatric intensive care. A week of clinical activity is described, with a focus on the interactions with computer systems that have become an integral part of patient-care activities for many of us. It becomes clear that the boundaries between personal computers, hospital systems, and the Internet are often not sharply defined. Resources that are used every week may include those residing on a personal digital assistant, on the hospital's electronic medical record, or on a distant site on the World Wide Web. Key resources on the Internet (World Wide Web and e-mail) are identified. The technical underpinnings, particularly the network that provides the infrastructure for various resources, are described.


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Pediatric heart failure management: solving the puzzle.

Wernovsky G, Hoffman TM.


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Completing the continuum of care. The growth of a pediatric hospice program.

Winkler WD, Mardegian CA.


This article examines the process of integrating a pediatric program into an existing hospice organization. It also discusses the five phases a child and family experience, from entering hospice through grieving the loss of the child.


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The impacts of distance to hospital on families with a child with a chronic condition.

Yantzi N, Rosenberg MW, Burke SO, Harrison MB.

*Social Science & Medicine* 2001;52(12):1777-1791.

Children with chronic conditions and their families face many similar challenges that can be stressful for the family including, daily caregiving activities, financial difficulties caused by unexpected expenses, and increased use of health services to treat and help manage the condition. Many of these families, in addition to facing daily caregiving...
responsibilities, must travel substantial distances to access some of the necessary aspects of their child's health care. In this study, the Burke et al, (1994-1996) data of repeatedly hospitalized children and their families are used to explore a geographical dimension of family impact, distance. Outcome measures from the Feetham Family Functioning Survey and the Questionnaire on Resources and Stress are analyzed using exploratory and multivariate analysis. Results show that distance to hospital plays a role in the two areas of family life regarding relationships within the immediate family, and issues surrounding the ability to maintain the child in the family home. The implications of the results for family, health care intervention, and government policies and guidelines are discussed.

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Living, not existing, beyond critical care.

Yu M.


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Advances in the diagnosis of cystic fibrosis in infants.

Zeitlin PL.


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Long-acting morphine for pain control in paediatric oncology.

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Background. Guidelines for treatment of paediatric cancer pain recommend the usage of long-acting morphine. However, published paediatric experience with this drug is restricted to 147 children not systematically evaluated, and thus insufficient. We aimed to systematically analyse the age-dependent effects and adverse effects of long-acting morphine in paediatric cancer patients. Procedure. Ninety-five children aged 1 to 19 years were enrolled in a collaborative retrospective study conducted over seven-and-a-half years. Pain was scored according to a numeric rating scale (NRS, range 0 to 5), and the corresponding medication was recorded. Results. In 83 children documentation period started during morphine treatment (71, oral long-acting; 1, rectal; 11, IV). Mean oral/equivalent morphine starting dose was 1.3 mg/kgbw/d (SD 0.9). Mean end dose was 2.8 mg/kgbw/d (SD 2.7). Infants aged < 7 years received the highest average dose (2.6 mg/kgbw/d, SD 2.8), while patients >12 years received the lowest dose (1.4 mg/kgbw/d, SD 1.1). Median pain intensity decreased from score 1.0 (mean 1.2) NRS at the beginning to 0 (mean 0.6) NRS at the end. The proportion of patients scoring > 2 NRS (severe or most severe pain) under morphine treatment decreased from 26 to 12% (P = 0.08). In children >12 years pruritus was frequently observed (23% of patients). In all age groups, there were no severe adverse effects during the study period.
Conclusions. In paediatric haematology/oncology, pain control by oral long-acting morphine proved to be safe and effective even in the very young patients. The pharmacological properties of long-acting morphine are ideally suited for paediatric use, combining efficacy and compatibility. (C) 2001 Wiley-Liss, Inc. [References: 22]

Unrelated umbilical cord stem cell transplantation for X-linked immunodeficiencies.


Banked unrelated umbilical cord blood matched at 5 of 6 human leukocyte antigen loci was used to reconstitute the immune system in 2 brothers with X-linked lymphoproliferative syndrome and 1 boy with X-linked hyperimmunoglobulin-M syndrome. Pretransplant cytoreduction and posttransplant graft-versus-host prophylaxis were given. Hematopoietic engraftment and correction of the genetic defects were documented by molecular techniques. Two years after transplantation, all 3 patients have normal immune systems. These reports support the wider use of banked partially matched cord blood for transplantation in primary immunodeficiencies.

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